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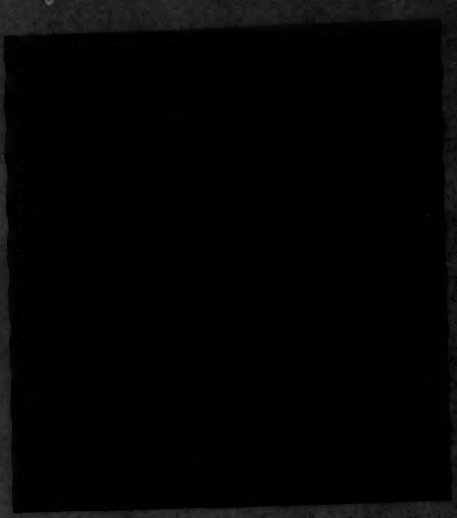

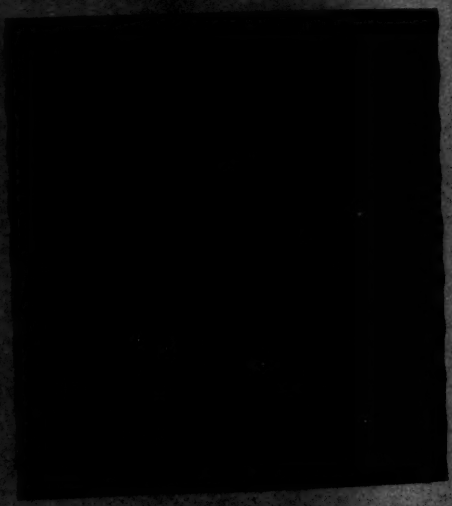

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OCTOBER, 1949

COMMUNICATIONS

STUDIES ON THE INTRA-OCULAR FLUIDS

Part 4 — The Dialysation of Aqueous Humour against Plasma*

BY

SIR STEWART DUKE-ELDER, HUGH DAVSON
and D. M. MAURICE

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THERE have been many attempts over the last quarter of a century to determine whether the aqueous humour and the blood plasma were in thermo-dynamical equilibrium. One of the most direct ways of determining whether or not this is so is to dialyse the one fluid against the other across a separating membrane; if there is a migration of substances across the membrane, it is obvious that such an equilibrium does not exist. In previous work of one of us (Duke-Elder, 1927) using pooled aqueous humour of

* Received for publication, March 14, 1949.

several animals, it was found that such a transfer was not of great dimensions; but the present experiments, carried out in much more efficiently controlled conditions, demonstrate that a transfer from the aqueous humour into the blood-plasma is consistently found.

METHODS

General.—Dogs and cats were anaesthetised with nembutal; aqueous humour was removed from both eyes with a gauge 1 needle and syringe; blood was obtained by heart-puncture, heparinised, and equilibrated with alveolar air at body temperature; the plasma was separated by centrifugation in a corked tube. For studies on the chloride content and on conductivity, the fluids from a single animal were sufficient for an experiment; for studies on the distribution of sodium, the fluids from three animals were pooled in strict proportion.

Dialysis.—In Experiments Nos. 1-12, the plasma was placed in a collodion sac, prepared as described by Dale and Evans (1920), tied to the end of a piece of capillary tubing (Fig. 1), which was then sealed with sealing-wax. The sac was washed with a few drops of aqueous humour and then immersed in sufficient of

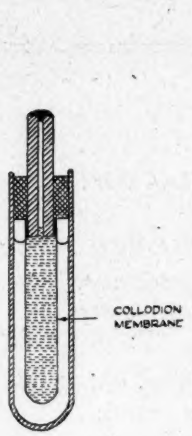

FIG. 1.

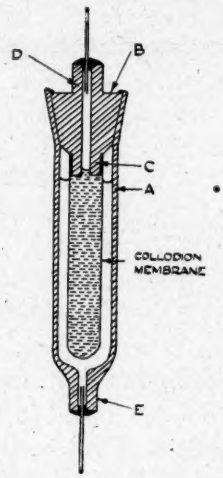

FIG. 2.

Dialysation apparatus

this fluid to cover it. A collar of rubber tubing round the capillary permitted the sac to stand in the fluid without touching the sides or bottom of the container. Evaporation was reduced (but apparently not always completely prevented) by covering the collar with paraffin-wax. The system was allowed to stand for about 16 hours at room temperature together with samples of the original fluids in stoppered tubes. During the course of the work it became evident that, in some of the experiments, evaporation of the fluids was taking place in the dialysing system; consequently the apparatus illustrated in Fig. 2 was devised. It consisted of an outer glass container, A, into which fitted a ground stopper, B. The base

of the stopper formed a slightly tapered projection, C, which was pushed into the top of the collodion sac, the junction being sealed with cellulose acetate paint. The stopper and the bottom of the container were pierced by holes, into each of which was sealed by sealing wax a length of glass tubing drawn out to capillary thinness. The stopper with its attached collodion membrane was placed in the container and fixed in position by wax or paint spread on the outside junction. An outer projection on the stopper, D, was pushed into a hole in a rubber bung; into the other end of this hole was pushed a glass tube joined to a length of rubber tubing so that by sucking or blowing on to this tubing the collodion sac could be collapsed or blown out. To fill the chamber, the sac was blown out, the bottom capillary placed under the required solution which was then drawn in by suction until the sac was completely collapsed. The chamber was then removed from the bung, inverted, and another similar projection, E, on the bottom of the container pushed into the hole. The sac was then filled with the other solution, normally plasma, by sucking it in through the capillary in the stopper. Some suction was maintained on the tube until the tip of this capillary was sealed with wax. The container was then removed from the bung and the other capillary likewise sealed.

By this means equal volumes of fluid were obtained inside and outside the sac. The sac was 8 mm. in diameter and the inside of the container about 11 mm. so that if the sac were cut so that it almost touched the bottom of the container, the fluid levels inside and outside were about the same. For the experiments described here two chambers were constructed to take 1 ml. or 2 ml. of solution inside the sac, but it is not of course necessary, although desirable, that the chamber should be completely filled either inside the sac or outside.

Stirring was achieved by fixing the chamber on a large pulley rotating at about 1 rev./sec.; at this speed the air bubbles in the compartments moved through the whole column of liquid and back during each cycle. Preliminary studies showed that equilibrium was established in about 30 min. but generally two to three hours were allowed.

To empty the sac the tips of the capillaries were broken off and the filling procedure followed in reverse. Even with the operations required in the repeated washing of the sac before filling, it should remain serviceable for at least half a dozen dialysations; it must, of course, be kept wet when not in use.

Chemical.—Sodium was determined by the Barber-Kolthoff (1928) technique on triplicate 0·5 ml. samples of the fluids, ashed as described in an earlier paper (Davson, 1939); the mean standard error of the thirty-four triplicate determinations was one part in 730; since the effects described in this paper involve changes of as much as one to two per cent. there is no doubt of their statistical significance. Chloride was determined on 0·2 ml. samples by the Sendroy technique (1937), the mean standard error of triplicate determination being one part in 300.

Dry weight.—About 1 ml. of plasma was evaporated to dryness on a sand-bath and heated for about 24 hours in an oven at 105°.

Conductivity.—The conductivity cell was in the form of a pipette of 0·3 ml. capacity with a flattened bulb into which were sealed two platinum wire electrodes, blackened in the usual manner. Connection to the bridge was made by two copper wires, soldered on to the electrodes and carried round the stem of the pipette. The junctions and leads were protected with a coating of wax and lacquer. The bulb was filled by suction through a rubber tube which was then clipped off and the tip of the pipette was sealed with a length of blocked cycle-valve tubing. The cell was immersed in melting ice contained in a Dewar flask. Before filling with a sample the cell could either be washed with distilled water and dried with alcohol and ether, or merely washed out with a fluid of conductivity close to that expected in the sample. The resistances were measured on a bridge constructed of normal radio components and calibrated with a decade resistance box. Duplicate readings on plasma or aqueous humour samples agreed to one part in a thousand.

pH.—The measurements were made with a glass-electrode reading against a calomel $\frac{1}{2}$ cell on a direct reading valve millivoltmeter. Alternate readings were taken of dialysed and undialysed fluids which were transferred as directly as possible to the measuring cell so as to avoid significant CO_2 loss.

RESULTS

Chloride.—The results with chloride on dogs are quite definite (Table I); as a result of a decrease in the original concentration of chloride in the aqueous humour (A_1), and of a rise in the corresponding concentration in the plasma (P_2), the ratio, R_{Cl}, changes from a mean value of 0·91 to one of 0·98. The two fluids are quite clearly not in equilibrium with respect to chloride. In cats the effects are not so large; nevertheless there is a definite

TABLE I

THE MOVEMENT OF CHLORIDES

The change in the Gibbs-Donnan ratio, R_{Cl}, following dialysis of plasma against aqueous humour. Expts. 21 to 24 refer to cats, the remainder to dogs. Expts. 1 to 16 were done with the earlier, 17 to 24 with the improved type of apparatus. A and P are the concentrations of chloride, in millimoles per kg. of water, in aqueous humour and plasma respectively. The suffices (1) and (2) refer to the figures obtained before and after dialysis respectively.

Expt.	A_1	P_1	R_1	A_2	P_2	R_2	$R_2 - R_1$
Dog 1	138.4	121.2	0.875	130.3	126.0	0.965	+ .09
2	145.4	130.0	0.895	140.1	134.6	0.96	+ .065
3	142.2	127.1	0.895	133.1	133.2	1.000	+ .105
4	134.5	123.9	0.92	129.0	129.9	1.007	+ .087
5	127.7	119.2	0.935	125.0	122.0	0.975	+ .04
6	131.7	123.0	0.935	132.1	128.5	0.975	+ .04
7	130.2	123.8	0.95	131.3	130.7	0.995	+ .045
17	128.0	118.2	0.923	126.9	123.0	0.97	+ .047
18	123.2	114.4	0.93	122.1	118.5	0.972	+ .042
19	129.9	115.9	0.893	121.6	122.4	1.005	+ .112
20	127.5	109.9	0 865	122.5	115.2	0.94	+ .075
Mean			**0.911**			**0.979**	
Cat 21	131.6	122.7	0.932	127.5	126.0	0.99	+ .058
22	127.5	123.9	0.973	128.9	124.6	0.968	— .005
23	130.6	119.1	0.91	129.5	123.4	0.953	+ 043
24	132.3	127.7	0.965	132.0	128.4	0.973	+ .C08
Mean			**0.945**			**0.971**	

change in the ratio following dialysis when this is low in the original fluids, but it would appear that in some animals the fluids are in equilibrium in respect to chloride whilst in others they are not. It will be noted that the decrease in concentration in the aqueous humour does not always equal the increase in plasma concentration, for the rise in concentration in the plasma was on occasions larger than the fall in the aqueous humour.

Sodium.—Some results with this ion are shown in Table II; they are equally consistent but not so striking in magnitude. The average value of R_{Na} changes from 1·036 to 1·057 in the dog,

TABLE II

THE MOVEMENT OF SODIUM

The change in the Gibbs-Donnan ratio, R_{Na}, following dialysis of plasma against aqueous humour. Expts. 12, 13 and 15 refer to cats, the remainder to dogs. Pooled specimens from three animals used in each experiment. A and P are the concentrations of sodium, in millimoles per kg. of water, in aqueous humour and plasma respectively. The suffices (1) and (2) refer to the figures obtained before and after dialysis respectively.

Expt.	A_1	P_1	R_1	A_2	P_2	R_2	$R_2 - R_1$
Dog 8	150.4	154.6	1.027	146.6	155.6	1.062	+ .035
9	150.3	155.0	1.032	147.4	154.7	1.050	+ .018
10	149.8	157.2	1.050	149.1	160.5	1.077	+ .027
11	148.5	155.8	1.049	149.1	158.7	1.064	+ .015
14	149.8	154.0	1.028	148.5	154.1	1.038	+ 01
16	147.4	152.1	1.032	145.7	152.8	1.049	+ .017
Mean			**1.036**			**1.057**	
Cat 12	155.8	160.2	1.028	156.6	166.9	1.066	+ .038
	155.8	160.2	1.028	158.5	168.2	1.061	+ .033
13	152.3	160.8	1.056	150.3	161.2	1.073	+ .016
15	152.1	158.7	1.043	149.3	159.3	1 067	+ .024
Mean			**1.042**			**1.068**	

and from 1·042 to 1·068 in the cat. Once again we find that the change in aqueous humour concentration is not always balanced by that in the plasma.

In Experiments 8 and 9 the sac had been maintained in distilled water before use; this produced a diluting effect on the system which obscured the rise in plasma concentration; in all succeeding experiments the sac was equilibrated with saline; there was, however, evidence of evaporation, an effect which tended to obscure the fall in concentration of the aqueous humour (Experiments 10-12). This is well brought out by Experiment 12; in this experiment sufficient fluids were obtained for two successive dialyses, and as a result of the first dialysis the value of R_{Na} rose from 1·028 to 1·066. New lots of the same fluids were then placed inside and outside the sac and the system left for another 16 hours; it will be seen that the value of R_{Na} did not differ significantly from that obtained after the first dialysis, but that as a result of evaporation the concentrations of sodium in aqueous humour and plasma both rose. In the later experiments (Nos. 13 to 16) evaporation was excluded by the use of the new type of chamber; R_{Na} invariably increased as a result of dialysis as before, but even under these conditions the loss in concentration in the aqueous humour was not entirely accounted for by a rise in the plasma concentration. It is not easy to explain

these discrepancies. It is possible that the collodion membrane itself is not in equilibrium with the fluids surrounding it; it may tend to turgesce, to adsorb or desorb ions, the particular effect observed depending on its previous history. In view of the large number of animals required and the extreme laboriousness of the chemical technique, it was not thought worthwhile investigating the point further; all ten dialyses, representing the fluid from 27 animals, concurred in showing that the ratio R_{Na} increased as a result of dialysis; dilution or concentration of the system by a few per cent. should produce slight errors in the ratio, but too small to be detected (cf. for example Expt. 12).

Conductivity.—The changes in conductivity are shown in Table III. In all cases there was an increase in the resistance of the aqueous humour and a decrease in that of the plasma, *i.e.*, the changes were such as could be produced by a movement of sodium

TABLE III

CONDUCTIVITY EXPERIMENTS

Changes in electrical resistance of plasma ($^{\Delta}R_{Pl}$) and aqueous humour ($^{\Delta}R_{Aq}$) as a result of dialysis. Expts. 25 to 33 on cats; Expt, 34 on a dog. In Expts. 30 and 32, whole blood was placed in the sac instead of plasma.

Expt.	$^{\Delta}R_{Aq}$	$^{\Delta}R_{Pl}$	$\frac{^{\Delta}R_{Pl}}{^{\Delta}R_{Aq}}$
Cat 25	1.7	−3.6	2.1
26	2.5	−8.7	3.5
27	0.6	−0.7	1.2
28	1.8	−2.1	1.2
29	1.0	−1.8	1.8
30	1.2	−2.6	1.45
31	7.3	−10.6	1.45
32	0.5	−1.7	2.4
33	0.1	−1.2	2.0
Dog 34	6.5	−4.3	1.5
Mean	**2.2**	**−3.7**	**1.7**

and chloride from the aqueous humour into the plasma. It may be computed, from the characteristics of the dialysis system, that the ratio: *Decrease in Resistance of Plasma/Increase in Resistance of Aqueous Humour* should be in the region of 1·4; the mean ratio is 1·7.

pH.—The increase in the value of R_{Na} might conceivably be due to an alkalinity of the dialysis system resulting from the loss of CO_2; the observed changes, however, would require an increase of about 50 per cent. in the base-binding power of the plasma, a change that could be achieved only by a change of pH in the region of one unit. It was not feasible to make pH measurements on the fluids used for analysis; separate dialysis

experiments were therefore carried out with both types of sac to determine whether any significant change in the pH of the two fluids took place as a result of the dialysis. It was found, in general, that the pH of the plasma was unaffected by dialysis whilst the aqueous humour tended to become more acid; presumably this is because the losses of CO_2 in the manipulation cause the aqueous humour to become alkaline; dialysis with the better buffered plasma tends to restore the pH to the physiological range.

The re-formed aqueous humour.—Earlier work (Davson and Weld, 1941) has shown that the aqueous humour re-formed after paracentesis is in close approximation to equilibrium with plasma

TABLE IV

THE SODIUM DISTRIBUTION IN PLASMOID AQUEOUS HUMOUR

The Gibbs-Donnan ratio, R_{Na}, in the re-formed aqueous humour. R_1 and R_2 are the ratios obtained with the normal aqueous humour before and after dialysis respectively; R_3 is the ratio obtained with normal plasma and re-formed aqueous humour.

Expt.	R_1	R_2	R_3	Protein in Re-formed Aqueous Humour
9	1.032	1.050	1.012	—
10	1.030	1.077	1.010	3 per cent.
11	1.049	1.064	1.024	2.5 per cent.
12	1.028	1.064	1.011	3 per cent

in respect to chloride: in the present work the fluid, re-formed within about half an hour of withdrawal of the normal aqueous humour, was analysed for sodium; the results are shown in Table IV. It will be observed that R_{Na} has become quite close to unity.

DISCUSSION

The results of these experiments show that the aqueous humour is not in thermo-dynamic equilibrium with blood plasma in so far as the distribution of sodium and chloride between the two fluids is concerned, for these substances are both in excess in the aqueous humour and migrate from this fluid to the plasma on dialysation. It might be suggested that the high concentration of sodium in the aqueous humour could be accounted for on the basis of a difference of pH between plasma and aqueous humour resulting, for example, from the formation of lactic acid in the metabolism of the lens; but such an explanation, even if it were acceptable, could not account for the fact that the excess of

chloride in the dog's aqueous humour actually exceeds that of sodium. The fact that arterial blood-plasma was arbitrarily chosen as the reference fluid may give rise to question, but the concentrations of sodium in arterial and venous plasma are not appreciably different, so that it is unlikely that the concentration in the capillary plasma—the fluid with which the comparison should be made—differs measurably from that in the arterial plasma used. We are therefore forced to the conclusion, already arrived at in our earlier chemical work, that energy is expended in transferring sodium and chloride from the blood to the aqueous, that is, that a process of secretion exists.

In the dog, the deviation of the concentration of chloride from equilibrium was considerably greater than that of sodium, the excess of chloride in the aqueous humour amounting to about seven per cent. or some 9 millimoles per litre; since the discrepancy of sodium amounts to only about 3 millimoles per litre, the results suggest that some other cation is present in excess; studies on the distribution of potassium (Davson, Duke-Elder and Benham, 1936), calcium and magnesium (Stary and Winternitz, 1932) have not revealed excesses of these ions in the aqueous humour of a magnitude sufficient to account for the excess of chloride. Presumably an organic cation is present. In the cat the deviation of the chloride ratio from the equilibrium value was considerably smaller, and on occasions it was found that the chloride ion was actually at equilibrium; in this respect, therefore, there seems to be a significant quantitative difference in physiology between the cat and dog.

The plasmoid aqueous humour formed after paracentesis is an entirely different case. It is seen that the ratio for the concentration of sodium (R_{Na}) as seen in Table IV approximates unity and this is most simply interpreted as indicating that during the rapid process of filtration associated with the re-filling of the anterior chamber, the diffusible ions filter through the blood-aqueous barrier in amounts proportional to their concentrations in the plasma. There is thus no attempt initially to form a Donnan equilibrium: only at a later stage would the resultant potential impose a different ionic distribution. This, of course, is in accord with the view that in the conditions of stress resulting from the collapse of compensatory pressure on the outer side of the capillary walls, the normal blood-aqueous barrier is disrupted and direct transference takes place through widely opened spaces between the endothelial cells so that a direct transudation of substances—and sometimes of whole blood—takes place.

SUMMARY

Direct dialysation of the aqueous humour against the blood plasma shows by chemical analyses and conductivity measurements that there is a transference of sodium and chloride from the former to the latter. The two fluids are therefore not in

equilibrium and in respect to sodium and chloride secretory energy must be postulated in the formation of the intra-ocular fluid.

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AN INVESTIGATION INTO THE MODE OF HEREDITY OF CONGENITAL AND JUVENILE CATARACTS*

BY

JOHAN SÆBØ

NORWAY

IT has been generally accepted since the investigations of Nettleship (1905) and Bateson (1909), that congenital and juvenile cataracts are inherited as dominant characteristics. It is reasonable that this should be so, for the literature provides many examples of cases in which such cataracts have been traced through several generations, and it leaves no doubt about the existence of dominant types.

There are, however, many reported cases of congenital and juvenile cataracts appearing sporadically, or in small numbers within the same family, and in which they have not been traced to succeeding generations. Such cataracts are frequently recorded as "familial"; they are often of the congenital or juvenile variety, and the possibility of their hereditary nature has received little consideration.

Franceschetti (1930) makes the following statement in Kurzes Handbuch der Ophthalmologie: "Das häufige isolierte Auftreten bei Geschwistern liesse auch an recessive Vererbung denken. Da aber Konsanguinität selten ist, kommt unregelmässige Dominanz in Frage."

Waardenburg (1932) says: "Auch die angeborene Katarakt wird im allgemeinen in dominanter Weise vererbt. Es liegen aber

*Received for publication, February 2, 1949

einige Beobachtungen vor, wodurch die Möglichkeit, dass es vielleicht eine rezessiv-erbliche Typen gibt, nicht ganz in Abrede gestellt werden kann. Die Rolle der Blutverwandtschaft ist eine geringe."

In "Handbuch der Erbkrankheitan" (1938), Bücklers has given the following declaration: "Das Schriftum ist zwar reich an Mitteilungen über erbliche Stare, doch sind viele Veröffentlichungen nicht zu verwerten, weil ausführliche Beschreibungen und gute Abbildungen fehlen. Es soll deshalb nicht verschwiegen werden, dass unsdr Wissen gerade auf diesem Gebiete noch lückenhaft ist."

Emphasis has been placed on the rôle of exogenous factors, *e.g.*, spasmophilia and rickets in the aetiology of just these types of cataract, and more recently attention has been drawn to the influence of virus diseases, *e.g.*, rubella. The possibility of hereditary influences tends to be overlooked and calls for further investigation.

With this possibility in view, all cases of congenital cataracts and juvenile cataracts which came under the author's care during six years as eye-specialist in *Telemark*, Norway, have been re-examined. This country district is relatively isolated and intermarriage is frequent, so that grouping of recessive genes is to be expected, with a correspondingly high number of homozygotes. Although statistical proof is lacking, hereditary eye diseases appear to be very common in this district.

Patients with congenital and juvenile cataracts were examined. In the published pedigrees the individuals physically examined are marked thus ◎. Information about their families was acquired from the patients themselves, their parents, their physicians, from parochial registers, and in some cases from other clinics.

The following are the more important points in the history of each patient:

Number of children in the family; number of affected individuals and their sex; convulsions or rickets in childhood; rubella or other diseases of the mother during pregnancy; consanguinity of the parents.

In the examination of the patient, the following points received attention:—

Type of cataract; comparison with types of cataract in affected siblings; uni- or bilateral; association with other defects or eye diseases, *e.g.*, nystagmus, microcornea; prognosis of the cataract.

The main object of this paper is to investigate the mode of inheritance in the reported cases of congenital and juvenile cataracts.

MATERIAL

(*a*) *Survey*

Within 17 families, 27 cases of congenital and juvenile cataracts are reported, 20 as congenital and 7 as juvenile cataracts. In 8 sibships the cataract was evident in 2 or more individuals. In 7 sibships consisting of more than 2 individuals, only 1 was affected. In 2 cases the affected person was the only child.

Among the 27 affected individuals, 14 were males and 13 females. In 9 of the 17 families the parents of the affected individuals were related, and in 5 of these the parents were first cousins. In 4 families the relationship was more distant.

As to the diagnosis, 20 cases are recorded as congenital cataracts, with different sub-titles — cat. totalis, subtotalis, nuclearis, membranacea, polaris posterior et polaris anterior. In 7 of the patients the diagnosis is cat. juvenilis, with the sub-titles of cat. corticalis posterior in 1 (Case 15) and cat. zonularis in the other 6. Possibly some of these zonular cataracts were, in fact, also congenital.

The cataracts were bilateral in all cases, except in Case No. 7. In Case No. 5 the cataract was unilateral at birth, but appeared later in the other eye.

Within 7 of these families (Nos. 1, 4, 5, 8, 12, 14 and 17) cases of cataracta senilis were also found, but only in 1 family (No. 12) was the senile cataract recorded in the father of patients with congenital cataract. In the rest, the senile cataracts were spread over other generations or within other side-branches.

Two of the patients (Cases Nos. 12 and 13) also suffered from retinitis pigmentosa, as well as cataracta corticalis posterior and polaris anterior, and in Case No. 13 surdo-mutitas was also found. One of the patients (No. 14) was feeble-minded.

In 2 of the patients (Nos. 22 and 23) there was a history of convulsions and rickets in childhood. Case No. 23 also had enamel defects of the teeth.

None of the patients' mothers could remember having had rubella or other infectious diseases during pregnancy.

Case Reports and Family Histories

Family 1: *Cases* 1 *and* 2.

O.S., male, born November 12, 1927, and A.S., male, born August 12, 1942, are No. 1 and 3 in a fraternity of three. Born in Bö, Telemark.

Case 1, *O.S.*, male. The cataract was noticed before the age of one year. Operation performed at age of 3 years in the right eye and at age of 3½ years in the left eye. A homogeneous greyish opacity covered all the pupillary area (3 mm. in diam.). No red reflex. Later on he was again admitted to a clinic for discissio cataractae o.u.

9*years old*: No nystagmus. Corneal diameter normal o.u. Visual acuity o.d. +13 = 5/15; o.s. +13 = 5/30. Cat. sec. o.u. The patient had always been

healthy. No convulsions. No indications of rickets. His mother was healthy during pregnancy.

Diagnosis: Cataracta congenita (subtotalis).

Case 2, *A.S.*, male. The patient's parents noticed the cataract immediately after birth. Otherwise he had been in good health. No convulsions. No indications of rickets. Mother healthy during pregnancy. Examined at age of 18 months and admitted to a clinic at age of 3 years.

3 *years old*: The patient was well-built and looked well. No sign of rickets. In both eyes there was an opacity in the central part of the lens and also considerable opacities in cortex. Discissio cataractae o.u.

5 *years old*: Repeated needlings. Vision: o.d. + 10 = 5/20; o.s. + 10 = 5/20.

Diagnosis: Cataracta congenita (subtotalis).

Pedigree information. See Fig. 1. The family was traced through four generations. In a fraternity of three (gen. IV) No. 1 and 3 had congenital cataract. No. 2, G.S., female, was normal (16 years old). The parents (G.S., male, 58 years, and

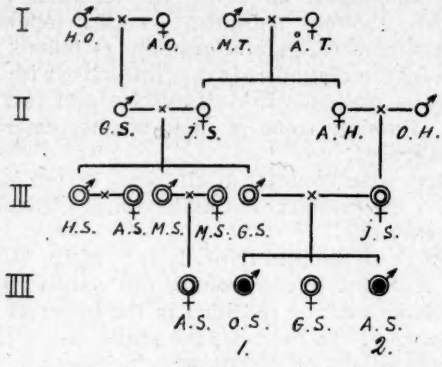

FIG. 1.

Family 1. Pedigree demonstrating that the parents of Case I, O.S., male, and Case II, A. S., male, both with congenital cataracts, were first cousins, H.O. (gen. 1) aged 76, and H. S. (gen. III) aged 64, had senile cataracts.

I.S., female, 47 years) of the affected children were normal. G.S., male, was No. 3 in a fraternity of three. No. 1, H.S., male, 64 years old, had a cat. senil. incip. with sector-shaped opacities in the anterior cortex. Vision: o.d. = 5/15; o.s. = 5/20. No. 2, M.S., male, 61 years old, his wife M.S., 62 years old, and their daughter, A.S., 17 years old, were all normal.

The grandparents of the affected children (O.S., T.S., A.H. and O.H., gen. II) all died at an advanced age (69, 79, 59 and 83 years) and had had good vision. Their great grandparents (M.T. and A.T., gen. I) died aged 45 and 34 years respectively, and had had good vision. The paternal great-grandfather (H.O., gen. I) had cat. senil. and died at the age of 76 years.

From the pedigree it is seen, that the patient's parents were first cousins.

Summing up, there were in this family two cases of cat. congen. The patient's parents were first cousins. Furthermore there were two cases of cat. senil. one in the patient's uncle and one in their great grandfather.

Family 2: *Case* 3.

M.D., female, born May 12 in Skien, and was No. 1 among two sisters.

Case 3, *M.D.*, female. History given by the parents of a healthy childhood. She had never had spasms or rickets. The cataract was noticed when 1 year old.

2 *years old*: Referred to a clinic for operation. At that time o.d.: Dense greyish

opacity in the pupillary area. No red reflex. O.s.: Grey opacity in the pupillary area with no shadow of the iris. She was operated on several times o.u. Attended elementary school and was able to read with the left eye. The right was almost blind. The patient's mother was said to have been healthy during pregnancy.

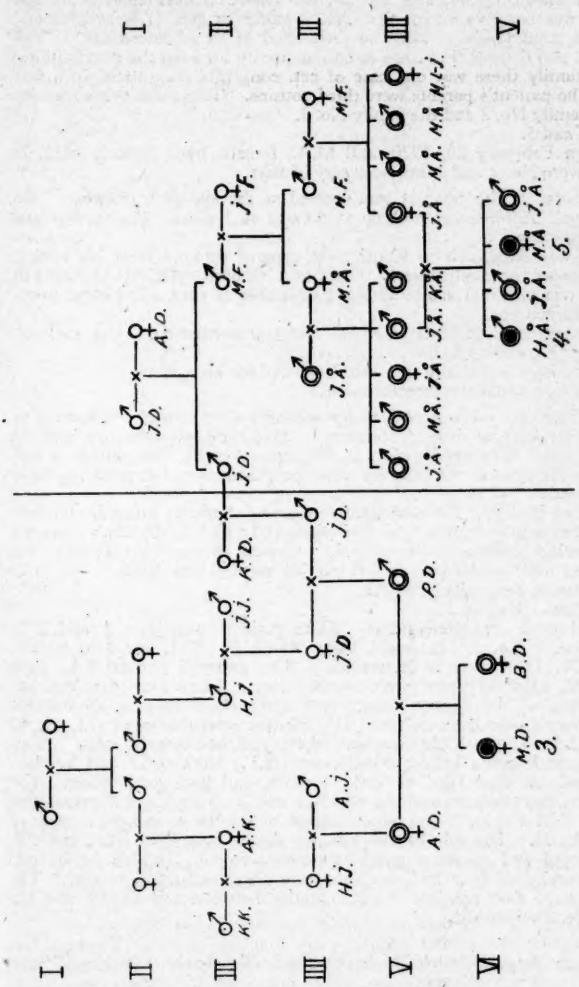

FIG. 3.

Family 3. Pedigree demonstrating that the parents of Case 4, H. A., male, and Case 5, M. A., female, both with congenital cataracts, were first cousins.

FIG. 2.

Family 2. Pedigree demonstrating that the parents of Case 3, M. D., female, with congenital cataract were third cousins. The consanguinity between the families 2 and 3 is also demonstrated.

21 *years old*: Nystagmus. Corneal diameter normal o.u. o. dext. in divergens. Total coloboma. Cat. sec. o.s. Pupillary area clear. Good red reflex. Vision o.d. P.L., O.s. = 5/10.

Diagnosis: Cataracta congenita (subtotalis).

Pedigree information. See Figs. 2 and 3. The family was traced through six

generations. The patient's sister (B.D., female, aged 22 years old) was normal. Their parents (A.D., female, aged 51 years, and P.D., male, aged 53 years old), were also normal. Their grand-parents (generation IV) died at the ages of 70, 39, 47 and 87 years, and had normal vision. Their great grand-parents (gen. III) died at the ages of 90, 90, 87 and 70 years, and had normal vision. Information about the other generations was negative except that No. 2 and 3 in gen. II were brothers, and that they had good vision. Both of them died at an advanced age. The pedigrees of No. 2 and 3 show that there is consanguinity between the two families.

Hence, in this family there was one case of cat. congenita (subtotalis) in a fraternity of two. The patient's parents were third cousins. There was consanguinity also between this family No. 2 and the family No. 3.

Family 3: *Cases* 4 *and* 5.

H.A., male, born February 20, 1920, and M.A., female, born June 2, 1922, in Siljan, Telemark, were No. 1 and 3 in a fraternity of four.

Case 4, *H.A.*, male. The cataract was noticed at the age of 6 months. No spasms. No rickets. Admitted to a clinic at the age of 3 years. His mother was healthy during pregnancy.

3 *years old*: It was impossible to identify the type of cataract from his record. Possibly it was a membraneous cataract. Operation on both eyes. At the ages of 4 and 5 years he was operated again, and was then able to read and passed intermediate school examinations.

17 *years old*: Nystagmus present. Corneal diameter normal o.u. Cat. sec. o.u. Vision o.d. $+8=5/20$; o.s. $+8=5/20$.

The patient was otherwise healthy and well developed for his age.

Diagnosis: Cataracta congenita (membranacea).

Case 5, *M.A.* The cataract was noticed immediately after birth. No spasms or rickets. Her mother healthy during pregnancy. Her right eye was operated on at the age of 2 years. Had good vision in left eye. Also in this patient it was impossible to state the type of the cataract from the description, but most probably it was a total cataract.

24 *years*: Looked healthy. No nystagmus. Corneal diameter normal. O. dext in divergens. The pupillary area was clear with red reflex. O. sin. Central opacity in posterior cortex. Vision o.d. Counts fingers at $\frac{1}{2}$ m.; o.s. $=5/20$. Operation still not advised in left eye, as reading was good.

Diagnosis: Cataracta congenita (totalis)?

Pedigree information. See Fig. 3.

Investigations through five generations. The patients were No. 1 and 3 in a fraternity of four. No. 2, J.A., male, born March 26, 1921, and No. 4, J.A., male, born July 30, 1923, were both normal. The patient's parents J.A., aged 56 years, and I.A., aged 55 years, were normal too. They were first cousins. The other members of the parents' group and their children were all normal. Three of the patient's grandparents (gen. III) died at advanced ages (72, 59, 82 years). No. 1, J.A., 92 years old, was still alive, and had normal eyes. Two of the patient's grandparents (M.A., female, and H.F.) were sister and brother. Their parents (gen. II) died aged 61 and 55 years, and had good vision. The connection between this pedigree and the previous one is obvious and demonstrates that the father of Case 3 (Fig. 2) was second cousin of the father and the mother of Cases 4 and 5 (Fig. 3). The side branches in the descendants after M.F. and I.F. (gen. II) were traced and no other cases of cataract found. In this family two patients of a fraternity of four had cat. congenita (?membranacea, totalis). The patient's parents were first cousins. Consanguinity between this family and the previous family No. 2 was noted.

Family 4: *Case* 6.

O.E., male, born August 26, 1939, in Rauland, Telemark, was No. 2 in a fraternity of two.

Case 6, *O.E.*, male. The cataract was noticed at the age of 1 year. His mother was healthy during pregnancy. He looked well. No spasms. No rickets.

1 *year old*: Nystagmus. Corneal diameter 9 mm. o.u.

In both of the lenses there were opacities, most dense in the central part, but also in cortex. Faint red reflex in the peripheral areas of the lenses.

5 years old: Mydriasis. o.u. The lenses were opaque all over, more dense in the central part than in the periphery. Faint red reflex from the lens-periphery. Vision: eyesight enough to walk by himself. The patient was operated on: Discissio et evacuatio cataractae o.u. Vision after operation difficult to state (+11=?5/20 o.u.).

Diagnosis: Cataracta congenita (totalis).

Pedigree information. See Fig. 4.

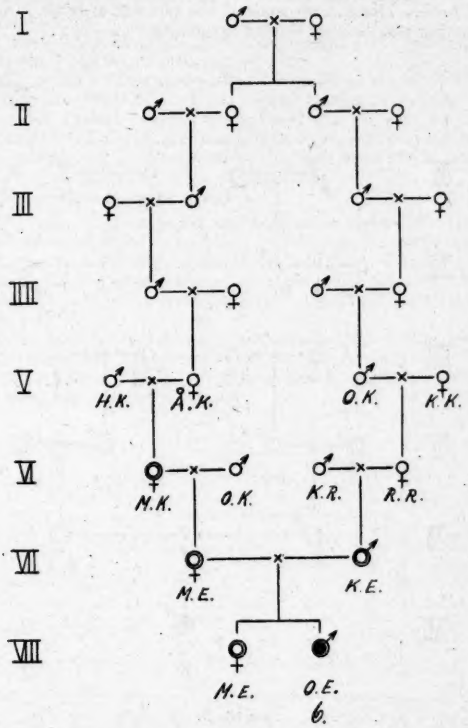

FIG. 4.

Family 4. Pedigree demonstrating that the parents of Case 6, O. E., male, with congenital cataract, were fifth cousins. O. K., gen. V, had senile cataract. (Aged 83).

The family was traced through eight generations. The patient's sister, M.E., was examined 18 years old, and was normal. His parents M.E., female, aged 49 years, and K.E., aged 68 years, were also normal. They were fifth cousins. The sisters and brothers of the patient's parents, and their children had normal eyes. The patient's grand-parents (gen. VI) of which only M.K., aged 78 years, was still alive, had good vision. Three of them died at advanced ages (70, 70 and 80 years old). From gen. V it was known that H.K., A.K., O.K. and K.K. died at advanced ages (81, 75, 83 and 90 years respectively) and, with the exception of O.K., who had cat. senilis, all had good vision. About the individuals in the other generations

I, II, III, and IV, it was only known that they died at advanced ages and probably had good vision. The descendency from gen. I was traced extensively, but no other cases of congenital or juvenile cataracts were found.

In this family, one patient in a fraternity of two had cat. congenita (totalis). The parents of the patient were fifth cousins. The patient's great grandfather had cat. senilis.

Family 5: *Case* 7.

I.L., male, born 1943, was the only child. Born in Mo, Telemark.

Case 7, *I.L.*, male. His parents noticed the cataract at birth. No spasms. No rickets. His mother was healthy during pregnancy.

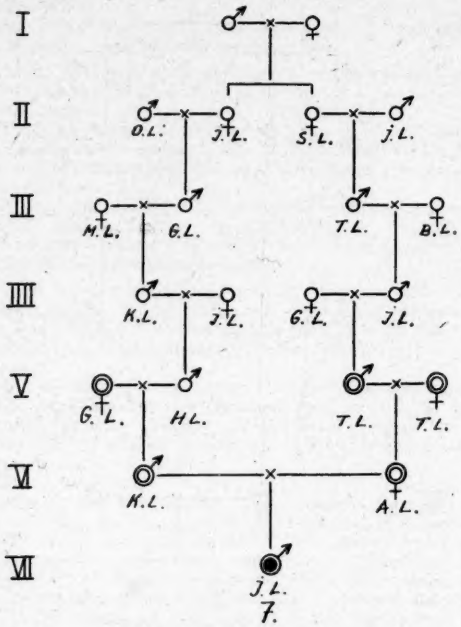

FIG. 5.

Family 5. Pedigree demonstrating that the parents of Case 7, J. L., male, with congenital cataract, were fourth cousins. G. L., gen. V, (aged 70), had senile cataract.

4 *years old*: The patient looked well. No nystagmus. Corneal-diameter normal o.u. *o.s.* In the central part of the lens there was a grey opacity formed as a disc. No riders. Red reflex around the opacity *o.d.* The lens was clear. Fundus normal. Vision: o.d. = C.F. at 1 m.: o.s. = 5/5.

Diagnosis: Cat. congen. (nuclearis) o.d.

Pedigree information. See Fig. 5.

Investigations into the family was carried out through seven generations. The patient's parents, K.L., male, aged 43 years, and A.L., female, aged 32 years, were normal. They were fourth cousins. The other members of the parents' fraternities and their children were normal. The patient's grandparents (gen. V) were still alive H.L. excepted, who died aged 58 years, and had good vision. The others

were G.L., female, aged 70 years, T.L., male, aged 62 years, T.L., female, 78 years old. The first one had cat. senilis with wedge-shaped opacities in cortex. Vision: o.d. = 5/20; o.s. = 5/15. The others had normal eyes. Information from the other generations disclosed no cases of cataract and these individuals died at advanced ages and were supposed to have had good vision. Investigations into their descendants disclosed no congenital or juvenile cataracts.

In this family, the only child of normal parents, who were fifth cousins, had cat. congen. (nuclearis). One of the grandmothers had senile cataract.

Family 6: *Cases* 8 *and* 9.

E.M., male, born May 31, 1901, and K.M., male, born June 28, 1905, in Mo, Telemark, were No. 1 and 3 in a fraternity of three.

Case 8, *E.M.*, male. The cataract was noticed at birth. Had always been healthy. No spasms or rickets. Mother healthy during pregnancy.

9 *years old*: He looked healthy. Nystagmus was present. Corneal-diameter normal o.u. Nuclear opacity in the lens, with a less dense zone around this. Good red reflex peripherally, o.u. Was operated in both eyes at the ages of 9 and 10 years, and was then able to read. Died 21 years old.

Diagnosis: Cat. congenita (nuclearis).

Case 9, *K.M.*, male. The cataract was noticed at birth. No spasms or rickets. Was 5 years old when admitted to a clinic. No disease in mother during pregnancy.

5 *years old*: Looked healthy. Nystagmus present. Corneal-diameter normal. In the pupillary area a greyish, 4 mm. diam. disc-like opacity. Red reflex peripherally. Probably a nuclear cataract o.u. Operated on both eyes. Vision o.d. + 11 = 6/36; o.s. + 11 = 6/36.

25 *years old*: In later years the vision had diminished. Glaucoma diagnosed o.d.

43 *years old*: Right eye became blind at the age of 26 years. Then glaucoma developed in the left eye. Vision: o.d. amaurosis: o.s. + 10 = 5/30. Intraocular tension o.s. 40 mm. Hg despite pilocarpine.

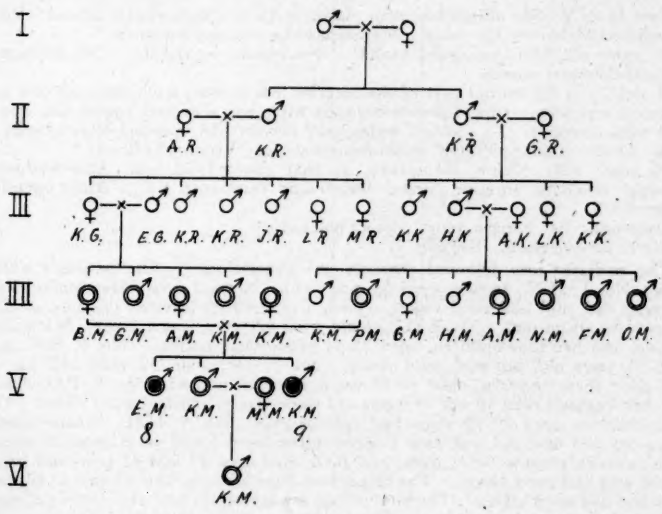

FIG. 6.

Family 6. Pedigree demonstrating that the parents of Case 8, E. M., male, and Case 9, K. M., male, both with congenital cataracts, were second cousins.

Diagnosis: Cat. congen. (nuclearis). Glaucoma.

Pedigree information. See Fig. 6.

Investigations through six generations were carried out. The patient's brother, K.M., aged 45 years, his wife, M.M., aged 28 years, and their child, K.M., 2 years old were normal. Their mother, B.M., aged 82 years, was No. 1 in a fraternity of five. All these and their children had normal eyes. The patient's father, P.M., aged 90 years, and No. 5, 6, 7 and 8 in his fraternity, and their children were all normal. No. 1, 3 and 4 of this fraternity died aged 86, 75 and 70 years old respectively, and had good vision. The paternal grandparents (gen. III) died aged 84 and 80 years, and had good vision. Three of their brothers and sisters died at an advanced age and had good eyesight. The maternal grandparents (gen. III), K.G. and E.G., died at the ages of 72 and 62 years and had good vision. In K.G.'s fraternity there were five other members who all died at advanced ages and had good eyesight. The patient's parents were second cousins.

In this family there were two cases of cat. congenita (nuclearis). The patient's parents were second cousins.

Family 7: *Cases* 10 *and* 11.

E.S., male, born 1885, and O.S., female, born 1889, in Siljan, Telemark, are No. 3 and 5 in a fraternity of nine.

Case 10, *E.S.*, male. He stated that he always had poor vision. Could read at school. His mother was healthy during pregnancy.

50 *years old*: He looked well and had always been healthy. No nystagmus. Corneal-diameter was normal o.u. O. dext.: In the central part of the anterior lens capsule there was a whitish opacity of 2·5 mm. diameter. In the posterior cortex there was a central rosette-like opacity of about 5 mm. diameter. Otherwise this lens was clear. O. sin.: The same picture. Fundus normal o.u. Vision: 5/20 o.u. No operation was advised, as ability to read was fair. He was drowned at age of 60 years.

Diagnosis: Cat. congenita (Cort. post. et pol. ant.).

Case 11, *O.S.* She always had poor vision, but was able to read at school. Vision diminished within the last years. Mother healthy during pregnancy.

50 *years old*: Enjoyed good health. No spasms or rickets. No nystagmus. Corneal-diameter normal.

O. dext. In the central part of the anterior lens capsule a whitish spot of 2 mm. diameter was seen. In the posterior cortex there was a central rosette-like opacity of 4 mm. diameter. In addition widespread powder-like opacities were seen in the lens. O. sin. similar picture. Fundus normal o.u. Vision=5/20 o.u.

59 *years old*: Vision diminished, at that time=5/30 o.u. Also wedge-like opacities noted in anterior cortex. Extractio cataractae o.d. After operation, vision=5/6.

Diagnosis: Cat. congen. (cort. post. et pol. ant.).

Pedigree investigation. See Fig. 7.

The pedigree was followed through five generations. The patient's siblings were: No. 1, H.N., female, aged 65 years old. Normal eyes. Her husband died 45 years old, and had good vision. Their five children between the ages of 22-46 years were all normal. No. 2, O.S., male, aged 62 years, was normal. No. 4, B.O., female, and her four children, aged 26-31 years, were normal. No. 6, N.S., male, died 41 years old, but had good vision. No. 7, J.S., male, 52 years old, his wife and their three children, aged 20-23 years, were all normal. No. 8, P.O., female, and her husband died 40 and 60 years old respectively, and had good vision. Their four children, aged 23-30 years had normal eyes. No. 9, H.H., female, aged 46 years old, her husband and their three children, aged 10-20 years, were all normal. The patient's parents N.O., male, and B.O., died aged 77 and 62 years old respectively, and had good vision. The father had three siblings who all died at advanced ages and had good vision. The mother had seven siblings who also died at advanced ages and had good eyesight. The patient's parents were first cousins. The patient's grandparents O.H., A.H., O.O., and E.O., died aged 89, 60, 70 and 80 years old respectively, and had good eyesight. (A.H. and O.O. were siblings.) The parents of the last mentioned (H.H., male, and J.H.) died aged 70 and 94 years old respectively, and had good vision.

The descendants from these were all traced as far as possible, but only the two recorded patients with cataract were found.

In this family two cases of cat. congenita. (cat. cort. post et pol. ant.) among nine siblings were recorded. The patient's parents were first cousins.

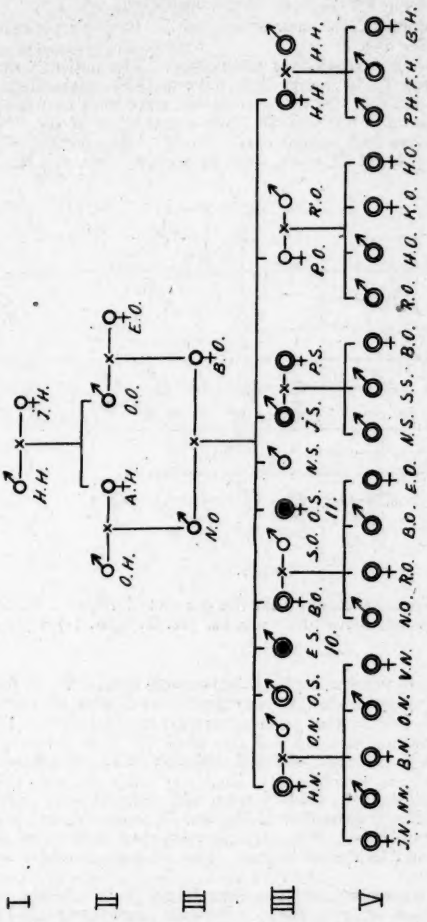

FIG. 7.

Family 7. Pedigree demonstrating that the parents of Case 10, E. S., male, and Case 11, O. S., female, both with congenital cataracts, were first cousins.

Family 8: *Case* 12.

K.K., male, born March 13, 1908, in Drangedal, Telemark, was No. 1 in a fraternity of five.

Case 12, *K.K.*, male. The patient was healthy during childhood. No spasms. No rickets. His eyesight had been poor as far back as he could remember. Difficulty in seeing in the darkness. His mother was healthy during pregnancy.

38 *years old*: General condition good. No nystagmus. Corneal-diameter normal o.u.

O. dext.: There was an anterior polar opacity of 2 mm. diameter in the lens. In the posterior cortex there was a rosette-like opacity. Otherwise the lens was clear. In the fundus retinitis pigmentosa was seen. O. sin.: Corresponding changes. Vision = 5/20 o.u. Visual fields were constricted.

Diagnosis: Cat. congenita (Cort. post. et pol. ant.). Retinitis pigmentosa.

Pedigree information. See Fig. 8.

The pedigree information covered four generations. The patient's siblings were normal. They were aged 27-35 years. His parents O.K., male, aged 64 years, and A.K., who died 65 years of age of tuberculosis, were both examined, and had normal eyes. The patient's father was No. 1 in a fraternity of six. No. 2, I.K., female, was unmarried, and had normal eyes. No. 3, A.M., female, 57 years old, and her three children, aged 24-28 years, were all normal. No. 4, J.K., male, aged

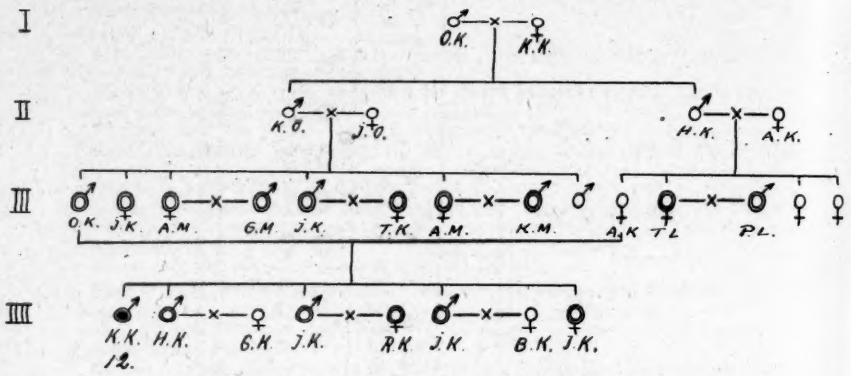

FIG. 8.

Family 8. Pedigree demonstrating that the parents of Case 12, K. K., male, with congenital cataract, were first cousins. K. O. (gen. 11), aged 71, had senile cataract.

54 years, had one child, 16 years old. Both had normal eyes. No. 5, A.M., female, aged 51 years, and her two children, 30 and 28 years old, were all normal. No. 6 died 20 years old, and had at that time apparently normal eyes. The patient's parents were first cousins.

The patient's mother had a sister who was still alive, T.L., 65 years old, and she had normal eyes.

His paternal grandfather, K.O., died 71 years old, and had then a senile cataract (from his physician). The grandmother, I.O., died 70 years old, and had good eyesight. His maternal grandfather, H.K., and grandmother, A.K., died aged 72 and 70 years respectively; and had good vision. The patient's paternal and maternal grandfathers were brothers.

In this family, one among five siblings were found to be affected. Diagnosis: Cat. congenita (cort. post. et pol. ant.). Retinitis pigmentosa. The patient's paternal grandfather had cat. senil. His parents were first cousins.

Family 9: *Case* 13.

A.K., female, born 1911, in Kragero, Telemark, and was No. 3 in a fraternity of three.

Case 13, *A.K.*, female. She had always had bad eyesight. She was deaf and dumb, otherwise healthy. She was well proportioned. Her mother had no disease during pregnancy.

37 *years old*: No nystagmus. Corneal-diameter normal o.u.

O. dext. There was a greyish opacity of about 3 mm. diameter in the anterior polar region of the lens. In the posterior cortex there was a rosette-like opacity. The nucleus of the lens was opaque. The fundus could not be seen.

O. sin. Corresponding picture.

It was difficult to obtain information about vision. The right eye was operated upon: Extractio cataractae. After operation retinitis pigmentosa was found. Vision: with +14 she was able to read big print.

Diagnosis: Cat. congenita (pol. ant. et cort. post.). Retinitis pigmentosa. Surdo-mutitas.

Pedigree information. See Fig. 9.

The pedigree information covered six generations. The patient was No. 3 in a fraternity of three. Her eldest sister, K.P., 50 years old, her husband, H.P., 60 years

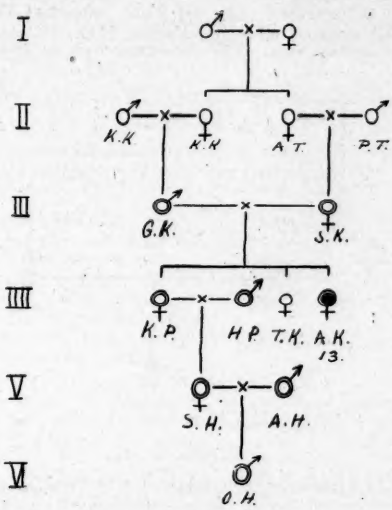

FIG. 9.

Family 9. Pedigree demonstrating that the parents of Case 13, A. K., female, with congenital cataract, were first cousins.

old, their daughter, S.H., 24 years old, and her husband, A.H. (28 years old) were all normal, together with their child, O.H., 4 years old. No. 2, T.K., female, died aged 31 years, but had good eyesight. The patient's parents, G.K., male, 70 years old, and S.K., 78 years old, had normal eyes. They were first cousins. The Patient's grandparents (gen. II) all died at advanced ages (64, 81, 84 and 83 years) and were said to have had good eyesight. Two of these, K.K. and A.T., were sisters. The descendants from these two were traced as far as possible and no other cases of cataract or other eye-disease were found.

In this family one sister among three had cat. congenita, (cat. pol. ant. et cort. post.). Retinitis pigmentosa and surdo-mutitas. The patient's parents were first cousins.

Family 10: *Case* 14.

S.B., female, born April 1, 1928, in Holla, Telemark. She was the only child.

Case 14, *S.B.*, female. During pregnancy the patient's mother had eclampsia. Spontaneous delivery. No infectious disease (*e.g.*, rubella). The mother noticed nystagmus in her child at birth. The child was admitted to a clinic 9 months old.

9 *months old*: Nystagmus. Total opacity of lens. Discissio cataractae o.u.
6 *months old*: Disc. cat. o.u.
9 *years old*: The patient was feeble minded. Not able to speak. Refused use of glasses. The pupillary-area was clear. Fundus appeared normal o.u. Nystagmus. Corneal diameter small (8½ mm.). Otherwise somatically normal.

Diagnosis: Cat. congen. (total). Imbecillitas.

Pedigree information. See Fig. 10.

The patient was the only child. Her parents, E.B., male, 45 years old, and K.B., 43 years old, were normal. The first one had four siblings, all normal. The second one had five siblings, all normal. There was no indication of relationship between the parents. The patient's paternal grandparents, R.B. and S.B., were both dead. The first one of pneumonia, 53 years old, and the second of apoplexia, 46 years old. Both of them had good eyesight. The patient's maternal grandparents were H.D., 80 years old, alive, with normal eyes, and M.D., who died 83 years old and had good eyesight. The paternal great-grandfather, H.D., 80 years old (gen. I) was still alive and had normal eyes. The other members of this generation died at

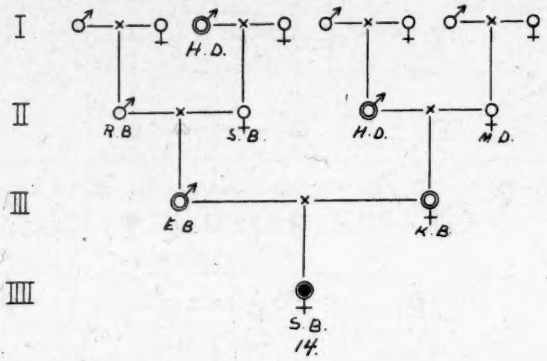

FIG. 10.

Family 10. Pedigree concerning Case 14, S. B., female, with congenital cataract. No consanguinity between the parents traced.

advanced ages and were supposed to have had good eyesight. The descendants from these were traced, but no other cases of eye-diseases were found.

In this family there was one case of cat. congenita (totalis) and Imbecillitas diagnosed. No indication of relationship between her parents.

Family 11: *Case* 15.

A.K., female, born May 2, 1924, in Gjerpen, Telemark, and was No. 2 of two siblings.

Case 15, *A.K.*, female. No indication of spasms or rickets in childhood. The patient's mother was healthy during pregnancy. For many years her eyesight was poor specially in bright day-light.

24 *years old*: Looked healthy. No nystagmus. Corneal diameter normal. There was a posterior cortical opacity in the lens. Otherwise the lens was clear. Fundus normal. The opacities were alike in both eyes. Vision: o.d = 5/10; o.s. = 5/10.

Diagnosis: Cat. juvenilis (cat. cort. post.).

Pedigree information. See Fig. 11.

Investigations through four generations were made. The patient's brother, K.K., 37 years old, his wife and their two children (aged 3 and 5 years) were all normal. Her parents, A.K., male, aged 67 years, and T.K., 70 years old, were both normal. Her father had two siblings, G.K., who died 74 years old, and K.K., 67 years old,

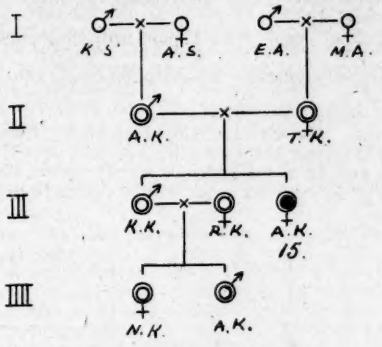

FIG. 11.

Family II. Pedigree concerning Case 15, A. K., female, with congenital cataract. No consanguinity between the parents traced.

both of which had good eyesight. Her mother had five siblings. Two of these died at advanced ages, but had good eyesight. Three were still alive, passed 60 years of age, and had normal eyes. Information indicated that the patient's grandparents died at advanced ages, and had good eyesight. The descendants were traced as far as possible, but no other eye-diseases were found. No consanguinity between the patient's parents was discovered.

One case of cat. juvenilis (cat. cort. post.) was diagnosed. No indication of relationship between the patient's parents.

Family 12: *Cases* 16, 17, 18, 19 *and* 20.

A.T., female, born April 24, 1903, I.T., female, born January 1, 1905, A.T., born June 30, 1907, S.T., male, born April 28, 1914, in Seljord, Telemark. They were No. 2, 3, 4 and 8 in a fraternity of eleven.

Case 16, *A.T.*, female. The cataract was noticed at birth. Had otherwise been healthy. No spasms or rickets in childhood. Able to read a little. The patient's mother was healthy during pregnancy. She was admitted to a clinic when 6 years old and then operated upon.

45 *years old*: Well proportioned and healthy looking. Nystagmus present. Corneal-diameter normal o.u. Left eye in divergens.

O. dext.: There was a dense secondary cataract in which there were two holes about 3 mm. in diam. Fundus appeared normal.

O. sin.: The pupil was peripheral, as a small coloboma. Fundus could not be seen. Vision: o.d. $+14=5/30$; o.s. $+14$ C.F. at 4 m. With $+14$ D lens she was able to read big print.

Case 17, *I.T.*, female. The cataract noticed at birth. Otherwise she had been healthy. No spasms or rickets. Her mother healthy during pregnancy. Operated on at 3 years old in the left eye and 7 years old in the right eye. Able to read and carry out housework.

43 *years old*: Nystagmus. Corneal diameter normal o.u. Well-proportioned and healthy-looking.

O. dext.: Anterior chamber deep. Pupil was round and dark. Fundus appeared normal.

O. sin.: Divergent strabismus. Anterior chamber deep. The pupil was peripheral as a small coloboma. There was a secondary cataract. Vision o.d. $+12=5/5$; o.s. $+12=5/20$.

Case 18, *A.T.*, female. The cataract noticed at birth. Otherwise of good health. No spasms or rickets. Her mother was healthy during pregnancy. The patient was operated on at 5 years old. Good vision post-operatively.

41 *years old*: Well-proportioned and looked healthy. Nystagmus present. Corneal-diameter normal o.u.

O. dext.: Ant. chamber deep. Round pupil with clear pupillary area. Fundus appeared normal.

O. sin.: Deep anterior chamber. Round pupil, with faint secondary cataract. Fundus looked normal.

Vision o.d. + 12 = 5/6; o.s. + 12 = 5/20.

Case 19, *S.T.*, male. The cataract noticed at birth. Otherwise healthy. No spasms or rickets. His mother healthy during pregnancy.

His eyes were operated on at the ages of 5 and 15 years. His eyesight remained poor, but he was able to read big print with a magnifying glass.

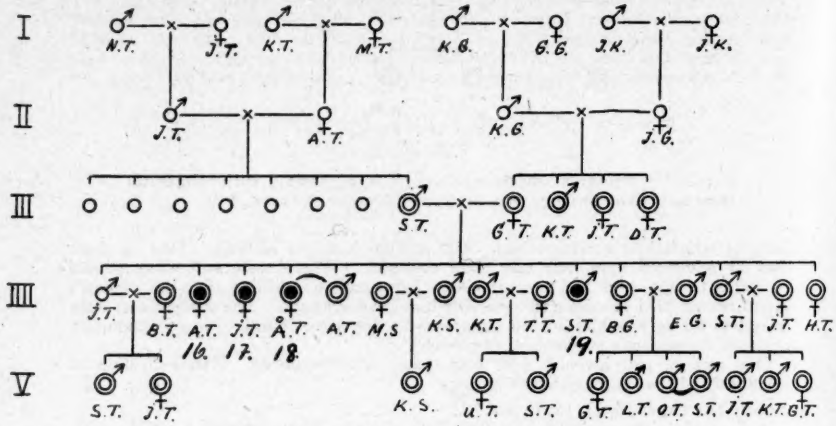

FIG. 12.

Family 12. Pedigree concerning Cases 16, A. T., female, 17, J. T., female, 18, A. T., female, 19, S. T., male, all with congenital cataracts. No consanguinity between the parents traced. S. T. (gen. III), aged 72, had senile cataract.

34 *years old*: He was well-proportioned and looked healthy. Nystagmus present. Corneal-diameter normal o.u.

O. dext.: Ant. chamber deep. Dense sec. cataract. Fundus was barely seen o.u.

Vision: o.d. + 12, count fingers at 3 m.; o.s. + 12 count fingers at 2 m. Discissio cataractae was advised.

Diagnosis: Cataractae congenitae.

Pedigree information. See Fig. 12.

The pedigree information covered five generations. The four patients had seven siblings, all normal. The patients were No. 2, 3, 4 and 8. No. 1 died 32 years old of tuberculosis. His wife and their two children (23 and 19 years old) were normal. No. 5, A.T. (twin of case 3) was normal. No. 6, M.S., 39 years old, and her husband and their children (4 years old) were normal. No. 7, K.T., his wife and their two children, aged 1 and 2 years respectively, were normal. No. 9, B.G., 33 years old, her husband, and their four children (twins), aged 6, 4 and 3 years, were normal. No. 10, S.T., 31 years old, his wife, and their three children aged 1 to 5 years, were normal. No. 11, H.T., 25 years old, was normal.

The patient's father, S.T., 72 years old, had cat. senil. incip. with wedge-shaped opacities in cortex.

Vision: o.d. = 5/6; o.s. = 5/10. Read easily. Seven of his siblings died between birth and 3 years. (Diphtheria, scarlatina, cholera, etc.) The patient's mother,

G.T., 68 years old, had normal eyes as also her three siblings, aged 66, 64 and 62 years. These three had thirteen children, all normal. The patient's paternal grandparents had good eyesight. The grandfather died 47 years old of pneumonia. The grandmother 38 years old of tuberculosis. The paternal great-grandparents died at the ages of 59, 91, 85 and 64 years, and had good vision. The maternal grandparents also had good eyesight. The grandfather died 64 years old (K.G.), and the grandmother 79 years old. The individuals in gen. I died all at advanced ages, 74, 64, 68 and 80 years old, and had good eyesight.

Investigations indicated no relationship between the patient's parents.

In this family four of eleven siblings had cat. congenita. Their father had cat. senil. incip. No consanguinity between the patient's parents was indicated.

Family 13: *Cases* 20 *and* 21.

T.H., female, born 1938, and O.H., male, born 1941, in Hjartdal, Telemark, were No. 2 and 4 in a fraternity of four.

Case 20, *T.H.*, female. The cataract was noticed before the age of one year. No spasms or rickets in childhood. The patient's mother was healthy during pregnancy. The patient's eyes were operated on at the ages of 4½ and 5½ years. Afterwards she had good eyesight.

9 *years old*: Well-proportioned and looked healthy. Corneal-diameter normal o.u. In both pupillary areas there was a fine membrane of sec. cataract. Discissio cat. o.u. was performed. Vision o.d. = 5/10; o.s. = 5/10.

No indication of the type of cataract was obtainable.

Diagnosis: Cat. congenita.

Case 21, *O.H.*, male. The cataract was noticed before the age of 1 year. He had always been healthy. No spasms. No indication of rickets. His mother was healthy during pregnancy.

4 *years old*: He looked healthy. Well-proportioned. Nystagmus present. The corneal-diameter normal o.u.

O dext.: In cycloplegia a central, nuclear opacity was seen in the lens, measuring about 5 mm. in diameter. No riders. Red reflex in the periphery.

O. sin.: The same picture. Vision: C.F. at 2 m. o.u.

Diagnosis: Cat. congenita (nuclearis).

Pedigree information. See Fig. 13.

The pedigree information covered three generations. The patients had two normal siblings, No. 1 and 3, 12 and 6 years old respectively. Their father, O.H., 40 years old, and his six siblings were all normal. Their mother, A.H., 35 years

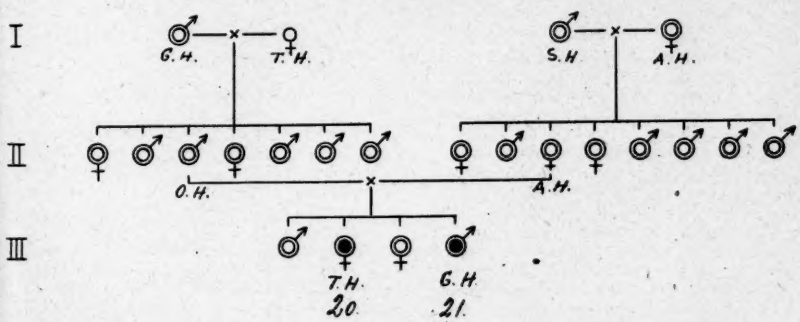

FIG. 13.

Family 13. Pedigree concerning Cases 20, T. H., female, and 21, G. H., male, both with congenital cataracts. No consanguinity between the parents traced. A sister of T. H.'s father (gen. I) had senile cataract. (Aged 65 years).

old, and her seven siblings were also normal. The paternal grandfather, G.H., 81 years old, was normal, and his wife, T.H., who died 60 years old, had good eyesight. A sister of the latter's father had cat. senilis, was operated on and regained good eyesight. The maternal grandparents, S.H., male, aged 64 years, and A.H., 64 years old, were still alive, and had normal eyes.

No indication of consanguinity between the patient's parents. The decendants from first generation were traced, but no evidence of cataract was noted.

In this family there are two cases of congenital cataract (nuclear) in a fraternity of four. No consanguinity between the patient's parents was indicated. One case of senile cataract far back in the family.

Family 14: *Case* 22.

A.S., female, born August 4, 1911, in Solum, Telemark. The patient was No. 7 in a fraternity of nine.

Case 22, *A.S.*, female. The patient's mother informed us that her daughter's eyesight was poor from childhood, and that she left school on that account at the age

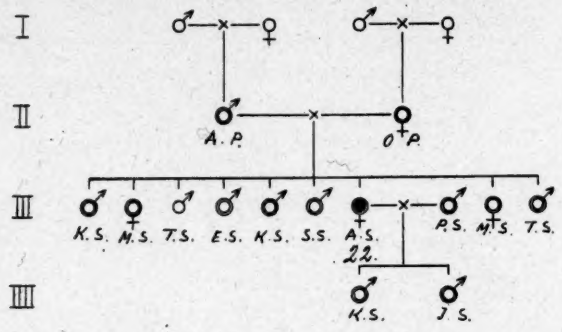

FIG. 14.

Family 14. Pedigree concerning Case 22, A. S , female, with juvenile cataract. No indication of relationship between her parents.

of 11 years. Spontaneously the mother also had asked the patient to state that she had spasms as a child and later also had rickets. The mother was healthy during pregnancy.

25 *years old*: She looked healthy and was well-proportioned. No nystagmus. Corneal diameter normal o.u.

O. dext.: In cycloplegia a typical perinuclear opacity of 5 mm. diameter, with riders, was seen. Red reflex around the opacity. *O. sin.*: same picture. Vision: Counting fingers at 3 m. o.u. Discissio et evacuatio cataractae o.u. was performed.

Vision: o.d. + 12 = 5/10; o.s. + 12 = 5/10.

Diagnosis: Cat. juvenilis (zonularis).

Pedigree information. See Fig. 14.

The pedigree information covered four generations. Her husband, P.S., was normal, as were their two children, aged 16 and 9 years. These had always been healthy, and were well-proportioned. The patient was No. 7 in a family of nine. No. 1 and 2 of these, aged 47 and 44 years respectively were normal. No. 3 died 8 days old. No. 4, 5 and 6 were 40, 38 and 36 years old, and were normal. So also were No. 8 and 9, 34 and 29 years old respectively. The patient's parents, A.P., male, aged 70 years, and O.P., 69 years old. were normal. The patient's grandparents died at advanced ages, and had good eyesight. There was no indication of relationship between the patient's parents. In addition to this information, the ascendants and descendants were followed as far as possible, and no other case of eye-disease was discovered.

In this family one patient with cat. juvenilis (zonularis) in a fraternity of nine was found. No consanguinity between her parents was indicated.

Family 15: *Case* 23.

I.K., male, born November 24, 1919, in Porsgrunn, Telemark, was No. 4 in a fraternity of five.

Case 23, *I.K.*, male. The patient's mother informed us that he had spasms as a nursling and later rickets. Was healthy later in childhood. Attended the elementary school, but had poor vision. His mother was healthy during pregnancy.

25 *years old*: Well-proportioned. Looked healthy. His teeth, however, demonstrated severe enamel defects. No nystagmus. Corneal diameter normal o.u.

O. dext.: Cycloplegia. A central perinuclear opacity of 5 mm. diameter in the lens was seen. Within this a lot of powdered opacities. Typical riders around the opacity. Fundus was normal.

O. sin.: Corresponding opacities. Vision: o.d. = 5/15; o.s. = 5/10. Able to read.

Diagnosis: Cataractae juvenilis (zonularis).

Pedigree information. See Fig. 15.

This pedigree information was followed through three generations. The patient's four siblings: No. 1, I.K., female, aged 39 years, No. 2, A.K., male, aged 35 years, No. 3, N.K., female, aged 32 years, and No. 5, B.K., female, aged 22 years, were

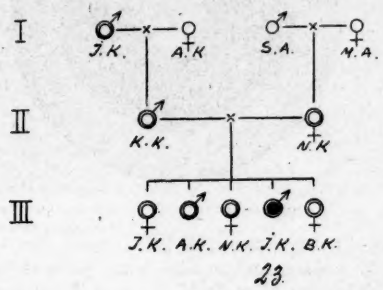

FIG. 15.

Family 15. Pedigree concerning Case 23, J. K., male, with juvenile cataract. No indication of relationship between his parents. J. K., gen. I, aged 91, had senile cataract.

all normal. They were all unmarried. The patient's parents (K.K., male, aged 64 years, and N.K., 64 years old) were normal. Of the patient's grandparents, only I.K., 91 years old, was still alive. He had cat. senil. incip. The other three died at advanced ages, 84, 82 and 87 years. They all had good eyesight. Their descendants were traced as far as possible and no other eye-diseases were discovered. No consanguinity between the patient's parents was found.

In this family there was one case of cat. juvenilis (zonularis), and one case of cat. senilis (in the paternal grandfather). No consanguinity between the patient's parents. The patient was said to have had spasms and rickets in childhood and demonstrated enamel defects of his teeth.

Family 16: *Cases* 24 *and* 25.

H.H., female, born April 11, 1927, and W.H., male, born 1925, in Brevik, Telemark, in a fraternity of two.

Case 24, *H.H.*, female. The patient had always been of good health. No spasms. No rickets. She had good eyesight until 18 years old, when she noticed blurring before her eyes, and became tired on reading. Her mother was healthy during pregnancy.

20 *years old*: Looked healthy and was well-proportioned. No nystagmus. Corneal-diameter normal o.u.

O. dext.: In the central part of the lens, there was a perinuclear opacity corresponding to the foetal nucleus. (See Fig. 16a.) Around this was a layer of fine powder-like opacities. In the periphery there were more riders. These riders were supposed to be located around the infantile nucleus.

Fundus appear normal. *O. sin.*: Corresponding opacities in the lens. Vision = 5/6 o.u.

Diagnosis: Cat. juvenilis (cat. zonular.).

Case 25, W.H., male. He had always been healthy. No spasms or rickets. Had good vision until recently, when he became tired on reading and noticed blurred vision at distance.

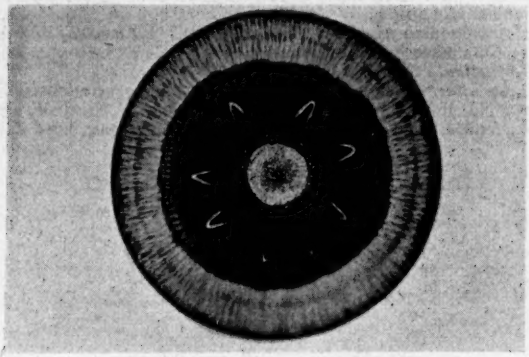

FIG. 16a.

Case 24, H. H., female, aged 20 years, with a particular type of zonular cataract. Her brother, Case 25, W. H., aged 16 years, demonstrates a similar cataract.

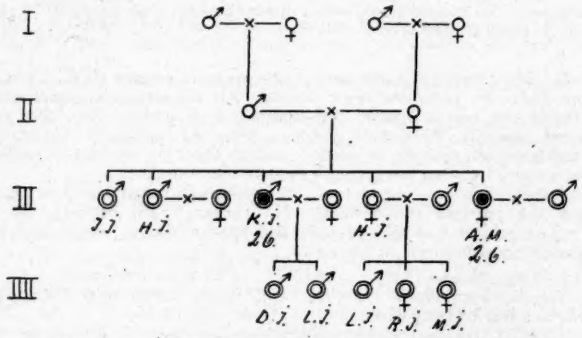

FIG. 16.

Family 16. Pedigree concerning Cases 24, H. H., female, and 25, W. H., male, with juvenile cataracts. No indications of relationship between their parents.

16 *years old*: He looked healthy. He was well-proportioned and had good teeth. No nystagmus. The corneal-diameter was normal o.u.

O. dext.: The clinical picture was here exactly the same as in his sister. There was a central perinuclear opacity with small powder-like opacities around this. Peripherally there were typical riders.

O. sin.: Similar opacities. Vision =5/6 o.u.

Diagnosis: Cat. juvenilis (zonularis).

Pedigree information. See Fig. 16.

The pedigree information covered three generations. The patient's parents, W.H., male, aged 52 years, and A.H., 46 years old, were both normal. There was no indication of relationship between these, as the first mentioned came from Sweden and the other from Norway. The patient's father had two elder brothers, who both died 62 and 57 years old respectively, one of tuberculosis and the other of heart disease. They had good eyesight. The patient's mother was No. 5 in a fraternity of seven. The other six were all examined. They were 60, 56, 53, 51, 38 and 36 years old and had normal eyes. Their children were also examined and demonstrated normal eyes. The patient's paternal grandparents died aged 67 and 84 years old. They had good eyesight. The maternal grandfather died 71 years old and had good vision. The maternal grandmother was still alive, 85 years old, and had normal eyes. The descendants from these were examined and no other cases of eye-diseases were found.

In this family both children in the family had exactly the same type of cataract, cat. juvenilis (zonularis). Consanguinity between the patient's parents was excluded. No indication of spasms or rickets in childhood.

Family 17: *Cases* 26 *and* 27.

K.J., male, born 1901, and A.M., female, born 1902, in Bamle, Telemark, were No. 3 and 5 in a fraternity of five.

Case 26, *K.J.*, male. He had always been healthy. No spasms. No rickets. He had always had poor eyesight. Able to read moderately well at school. From the age of 25 years he was convinced that something was wrong with his eyes. His mother was healthy during pregnancy.

36 *years old*: Looked healthy and was well-proportioned. No nystagmus. Corneal diameter normal o.u.

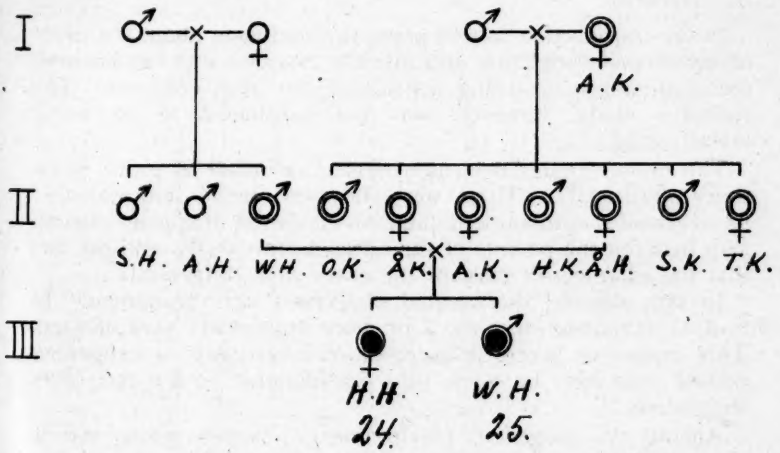

FIG. 17.

Family 17. Pedigree concerning the Cases 26, K. J., male, and 27, A. M., female, with juvenile cataracts. No indications of relationships between their parents.

O. dext.: Cycloplegia. A central perinuclear opacity of 5 mm. diameter. Some riders present. Red reflex around the opacity. Fundus appeared normal.

O. sin.: Corresponding opacity. Vision: o.d. +3=5/20; o.s. +3=5/15.

42 years old: The opacity more dense. Vision: +3=5/50 o.u. Extractio cataractae performed on both eyes. Vision: o.d.=5/15; o.s.=5/10.

Diagnosis: Cat. juvenilis (zonularis).

Case 27, A.M., female. She had always been healthy. No spasms. No rickets. Poor vision from the age of 17 years, specially in bright light. Her mother was healthy during pregnancy.

38 years old: Looked healthy and was well-proportioned. No nystagmus. Corneal diameter normal o.u.

O.U.: The lens opacities showed the same picture as in the above brother. Fundus normal. Vision: −1=5/15 o.u. Extractio cataractae o.u. was performed. Vision: o.d. +11=5/10; o.s. +11=5/10.

Diagnosis: Cat. juvenil. (zonularis).

Pedigree information. See Fig. 17.

The pedigree data covered four generations. The patients were No. 3 and 5 in a fraternity of five. No. 1, J.J., male, aged 57 years, No. 2, H.J., male, aged 50 years, and his wife, 48 years old, had normal eyes. Case 26, K.F., had a wife 45 years old and two children, D.J., male, aged 25 years, and L.J., male, aged 14 years, all with normal eyes. No. 4, H.J., female, aged 42 years, her husband, 36 years old, and their three children, aged 13, 17 and 19 years, were all normal. No. 5 was the patient, Case 27, who was married, but had no children. The patient's parents died 80 and 77 years old and had good eyesight. Information about the patient's grandparents indicated no eye-disease. They died at advanced ages, and probably had good eyesight.

In this family two cases of cat. juvenilis (zonularis) were diagnosed in a fraternity of five. No indication of relationship between the patient's parents. No indication of spasms or rickets during the patient's childhood.

DISCUSSION AND CONCLUSION

A. Heredity

In the introduction to this paper the confusion about the mode of heredity of congenital and juvenile cataracts was emphasized, the dominant mode being considered the most common. The recessive mode, however, was not considered to be worth investigating.

The present material is numerically too small to prove recessivity statistically. There were, however, many indications of its existence: pronounced familial incidence, frequent relationship between the parents of the affected individuals, and the fact that the cataracts in most of the cases were congenital.

In this material the familial incidences were prominent. In 8 of 17 examined sibships, 2 or more individuals were affected. This cannot be accepted as proof of inheritance — exogenous factors must also be taken into consideration — but it is very suggestive.

Among the exogenous (environmental) factors giving rise to congenital cataracts, special attention during recent years has been paid to the virus diseases (rubella). A considerable literature already exists on this subject, and a rather complete

list is presented by Bardram and Brändstrup (1947). There is, of course, no doubt that rubella in the mother during pregnancy may cause a congenital cataract, but in the cases presented this etiological factor can be excluded.

From the available literature in this field, it appears that the whole clinical picture caused by rubella is different from the clinical picture of the above cases. The cataracts caused by rubella usually are accompanied by considerable somatic defects and malformations, and hence must be considered a more serious disease. It is also evident from the present material that rubella cannot be made responsible for many congenital cataracts within the same sibship.

In the Table I below, the frequency of relationship between the parents of the affected individuals is demonstrated. In 5 of the 17 families the parents were first cousins. In another 4 they were related further back. Consanguinity was found, therefore, in 9 of the 17 families. Taking into consideration the remarks made in the introduction about the rare occurence of consanguinity, these facts are of great interest, and may indicate recessive heredity.

TABLE I

Number of children, number of cataracts, application of Weinberg's methods, and intermarriages in 17 families with congenital and juvenile cataracts.

Fam. No.	No. of Children (S)	No. of Catar. (T)	Weinberg's Meth. T (S—1)	Weinberg's Meth. T (T—1)	Intermarriages in Parents
1	3	2	4	2	First cousins
2	2	1	1	0	Third cousins
3	4	2	6	2	First cousins
4	2	1	1	0	Fifth cousins
5	1	1	0	0	Fourth cousins
6	3	2	4	2	Second cousins
7	9	2	16	2	First cousins
8	5	1	4	0	First cousins
9	3	1	2	0	First cousins
10	1	1	0	0	No
11	2	1	1	0	No
12	11	4	40	12	No
13	4	2	6	2	No
14	8	1	7	0	No
15	5	1	4	0	No
16	2	2	2	2	No
17	5	2	8	2	No
Total:	70	27	106	26	

Unfortunately, the present cases are too few for statistical proof as to the frequency of first-cousin marriages. (Only first-cousin marriages are used for this purpose, as only information on the normal frequency of these is available.) In this survey the frequency is 29·4 per cent. Normally it is less than 1 per cent. among the West European population (Dahlberg, Dunn).

Special interest should be paid to the pedigrees belonging to the families Nos. 2 and 3 (See Figs. 2 and 3). The parents of the patient, Case 3, were third cousins, and the parents of the Cases 4 and 5 (family No. 3) were first cousins. There were also relationships between the two families, as the father of Case 3 was second cousin to both the father and the mother of patients Nos. 4 and 5. In both of these families the cataracts appear to have been of the same type. They were all congenital and were operated on at the same age (3, 2, 3 and 2 years, respectively). Hence, they must be considered genotypical.

Among 70 siblings 27 are affected. This represents 38 per cent. of the total number, and indicates recessivity, but the number affected was higher than expected from the Mendelian rules for mono-recessive heredity: Normal: affected = 3 : 1. The material here is, however, a selected one, and a so-called recessive-excess is to be expected.

If Weinberg's sibling-method is applied to this material (see Table I), the following results are obtained: The percentage of affected children then is $100\frac{t\,(t\text{-}1)}{t\,(s\text{-}1)}$. S. is the number of siblings in a family, t is the number of affected children. t (s-1) is the siblings of affected children.* The result is then 24·5 per cent., which is in accordance with expectations when mono-recessive inheritance is involved.

As mentioned, 20 of the 27 cases were recorded as congenital. Possibly some of the cases recorded as juvenile were also congenital.

The cataracts described were not sex-linked, as 14 of the affected individuals were males and 13 females. They must therefore be considered autosomal.

The material shows no evidence supporting the hypothesis that the genes are incompletely dominant.

As the parents of the affected individuals, with only one exception, were normal, and the cataracts for the greater part congenital, the familial incidence high, and the consanguinity among the parents of affected individuals frequent, the mode of heredity was probably autosomal recessive in nature.

* t (t-1) is the affected siblings of affected children.

In 7 of the families (Nos. 1, 4, 5, 8, 12, 13 and 15) cat. senilis, as well as cat. congenita and juvenilis, was demonstrated. In one of these families, the father of 4 patients with congenital cataracts had senile cataract. Otherwise the senile cataracts were found in other generations or within side-branches. The patients with senile cataracts were all of considerable ages (76, 64, 83, 70, 71, 72, 65 and 91 years).

In the above-mentioned family (No. 12) the father of the 4 patients had an incipient senile cataract with wedge-shaped opacities in the anterior cortical layers of the lens. Visual acuity (72 years old) was 5/6 o.d.; 5/10 o.s. This cataract, accordingly, was entirely different from the congenital, total cataract in his 4 children.

The phenomenon of anticipation, in which cataracts appear at a younger age in every new generation, might explain the appearance of senile, juvenile or congenital cataracts within the same family. But this conception receives no support from the present investigations. The senile cataracts, Case No. 15 excepted, were all in families in which the patients had congenital cataracts. In cases of " Anticipation," intermediate types would be expected. The appearance of senile cataracts within these families therefore suggests a normal occurrence within the population.

In the material dealt with in the present paper there were 4 families (Nos. 14, 15, 16 and 17), representing 6 cases (Nos. 22, 23, 24, 25, 26 and 27), with zonular cataracts. It is mainly in this type that spasmophilia and rickets are supposed to be of etiological interest. This question, however, is not as yet solved. Franceschetti mentions that dominant inheritance is preponderant in zonular cataracts, but that recessive types (Baker, 1892, and Starbeck, 1877) also have been recorded. This writer also mentions spasmophilia and rickets as etiological factors.

In " Kurzes Handbuch der Ophthalmologie," Jess says (1930): " Nach allem ergibt sich dass eine einheitliche Schichtstargenese nicht angenommen werden kann."

In the present material, 2 patients (Nos. 22 and 23) among 6 suffered from spasmophilia and rickets in childhood. One of these (No. 23) demonstrated also considerable defects in the enamel of his teeth. In the other 4 patients with zonular cataracts (families Nos. 16 and 17) there was familial incidence, but neither the case histories nor the objective findings indicated spasmophilia or rickets.

The clinical picture of the 2 patients (Nos. 24 and 25 in family 16) is of great interest, as the zonular cataracts demonstrated peculiar arrangement of the lens opacities (see Fig. 16*a*, p. 26). The perinuclear central opacity probably is located in the foetal

nucleus, which develops between the 3rd and 8th months of foetal life. Peripherally in the lens some riders were seen. These riders are supposed to be located in the area of the infantile nucleus, which develops between the last weeks of foetal life and puberty. These findings may give the impression that some agents have been active at different periods. It does not disprove, however, that these cataracts may be due to genes.

In the present cases of zonular cataracts dominant mode of heredity is not indicated. Recessivity may be present, but cannot be proven. Neither can the effect of spasmophilia or rickets be assessed. Even if the aetiology of zonular cataracts is more obscure than that of other congenital or juvenile cataracts recorded in this paper, they are—with reservation—listed together in a common group.

As the patients were treated surgically in different clinics, and descriptions of the various types were often lacking, the differential diagnosis was rather difficult to make. With these reservations in mind, it would seem that the cataracts were of the same type within the same family. Thus, the type was considered to be similar (cat. subtotalis) in patients Nos. 1 and 2 in family No. 1. The cataracts were congenital and the two patients were both operated on at the same age (3 years). In the families Nos. 2 and 3, which were related, the cataracts also were congenital, and the patients were operated on at the same ages (2, 3 and 2 years). The diagnoses made were: cat. subtotalis, cat. membranacea and cat. totalis. In family No. 6 the diagnoses in Cases Nos. 8 and 9 were cat. nuclearis. Both were congenital and were operated on at the age of 9 and 5 years, respectively. In family No. 7 (Cases No. 10 and 11) the diagnoses were cat. corticalis and polaris ant., and the cataracts were of the same type. In family No. 12 (Cases Nos. 16, 17, 18 and 19) no diagnosis was made as to the special type, but they were all considered to be the same, as they were all congenital, and were operated on at the same ages (6, 3, 5 and 5 years, respectively).

In family No. 13 (Cases Nos. 20 and 21) the diagnosis was cat. nuclearis. Both of them had congenital cataracts. One of the patients was operated on when 4 years old, the other was ready for operation at a corresponding age. In family No. 16 (Cases Nos. 24 and 25) the zonular cataract was precisely of the same type in both brother and sister. In family No. 17 (Cases Nos. 26 and 27) there was a zonular cataract of the same type in both sister and brother. These cataracts were diagnosed and operated on at the same age. The type was, however, quite different from the one in the previous cases. Accordingly, the present

cases show that the types of cataracts vary from family to family, but within the same family the congenital or juvenile cataracts are much alike. These observations support the above-mentioned hypothesis that these cataracts are probably due to genes, and that there is no genetical connection established between these and the sporadic senile cataracts occurring in these families.

B. Occurrence of other diseases or symptoms in cataract patients

The occurrence of spasmophilia, rickets and enamel defects of the teeth has already been mentioned.

In one of the patients (Case No. 14) glaucoma was diagnosed. In two of them (Cases Nos. 12 and 13) the cat. corticalis posterior et polaris anterior was combined with retinitis pigmentosa. Case No. 13 also demonstrated surdo-mutitas.

As to the connection between the cataract and retinitis pigmentosa, two possibilities exist:

1. The cataract is correlated to retinitis pigmentosa.
2. The cataract is secondary to retinitis pigmentosa.

It is generally accepted that retinitis pigmentosa is as a rule a recessive condition. Kjerrumgard (1948) has found that 9 per cent. of all patients with retinitis pigmentosa demonstrated cat. pol. post., and that 5 per cent. of them had other types of cataract. He also demonstrated that 15 per cent. of the patients with retinitis pigmentosa had surdo-mutitas.

In our two cases of retinitis pigmentosa and cat. pol. post. there was also a cat. pol. ant., a type usually considered to be congenital. It is therefore reasonable to suggest that cat. pol. post., cat. pol. ant., retinitis pigmentosa and surdo-mutitas are correlated and that they are due to recessive genes.

Nystagmus was noted in 12 of the 27 patients and occurred only when combined with congenital cataracts.

Microphthalmos was found in 2 patients (Cases Nos. 6 and 14). Minor variations in the corneal diameters may have been more frequent, but exact measurements were not undertaken.

C. The prognosis

The results of the cataract operations were good, except in the following patients: Case No. 21, who was not operated upon; Case No. 14, who was feeble-minded; Case No. 9, with glaucoma; Case No. 19, with cataracta secundaria; Cases Nos. 12 and 13, with retinitis pigmentosa. Most of the operated patients were able to read and do full-time work in their different occupations.

In these conditions, therefore, the only advice to be given from

the eugenic point of view is to warn members of the families against inter-marriages. But as already noted, dominant types of congenital cataracts are likely to exist, and the mode of heredity in each case must be thoroughly investigated.

D. Conclusion

In the introduction it is shown how uncertain is the etiology and pathogenesis of congenital and juvenile cataracts. The dominant mode of inheritance is preponderant, and the recessive type of cataract is relatively rare.

Even if the material dealt with in this paper is too small to give proof by statistics, a series of cases of congenital and juvenile cataracts is cited which seems to indicate recessive inheritance.

Summary

The author of this paper has examined 17 families in which congenital or juvenile cataract occurred. Special attention was paid to their aetiology and mode of heredity.

In the introduction a review of the previous results from literature in this field was given. This showed that the dominant mode of inheritance is supposed to be the most frequent, with relatively rare examples of recessive inheritance. In recent years much attention has been paid to the virus diseases as causes of these cataracts, and inheritance has receded more into the background.

Exogenous factors, as spasmophilia and rickets, were suggested causes of zonular cataracts, but the rôle of heredity was also emphasized in this type.

Following this introduction, a review is given of the present material and the case reports with pedigrees.

In all, 17 families with 27 cases of congenital or juvenile cataracts have been examined. From these, 20 were recorded as congenital and 7 as juvenile. In each of 8 of these families, 2 or more individuals were affected. Among the 27 affected, 14 were males and 13 females.

In 9 of the 17 families, relationship between the parents of the affected individuals existed, and in 5 of them the parents were first cousins. In the other 4 families the relationship was less close.

In a following chapter, "Discussion and Conclusion," the results of these investigations were stated. Among the introductory remarks it was pointed out that the present material is too small to prove statistically the probable mode of inheritance in the cases investigated in this survey. As a eugenic measure,

the members of these families must be warned against intermarriage, but, as dominant types also occur, it is emphasized that the mode of inheritance must be studied in each case.

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THE MOTOR IMPULSE ELICITED BY THE RETINAL STIMULUS AND THE BINOCULAR OPTICAL REFLEXES*

BY

J. E. WINKELMAN

AMSTERDAM

IN order to study the problem of strabismus, it is necessary to investigate the disturbances of the normal binocular reflexes which occur in such patients. To do this, however, an exact knowledge of the essential features of these binocular reflexes in normal persons is required.

We started from the fundamental supposition that stimulation of a certain part of the retina not only produces a sensory effect, but is also active in a motor sense. In our opinion stimulation of the retina gives rise to a sensory *and* a motor impulse. There is evidence that in general these motor impulses may produce the following four physiologic phenomena:

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Objectively:

I. A sidewards movement of equal amplitude and in the same direction of both eyes; a conjugate movement.

II. A movement of the eyes in opposite direction: disjunctive movement; this results in a fusional movement.

Subjectively (when the motor effect on the muscles is inhibited).

III. Localisation of the stimulus in space.

IV. The visual sensation of apparent movement.

We have investigated three of the four reactions (II, III and IV). In the course of our experiments it appeared that in haploscopic alternating stimulation of both retinas with a small luminous spot we had an excellent method for analyzing these reactions.

DESCRIPTION OF THE APPARATUS.

A narrow strip of wood was fixed on a wooden frame, forming part of the circumference of a circle. Two wooden boxes may be shifted along this strip. Each box is provided with a small hole of 3 mm. diameter in the centre. These holes are covered by transparent paper and are illuminated by a $3\frac{1}{2}$ volt lamp within the boxes. An alternating device was incorporated in the electrical circuit; this makes it possible to illuminate the boxes alternately. The alternating device is driven by a motor (M) with adjustable speed; by lowering or increasing the speed of the motor the frequency of alternation can be controlled. The latter is measured by a revolution-counter (R), which may be placed on the axis of the alternating device (A). The experiments were carried out in complete darkness. The image of the luminous spot intended for the one eye was screened from the other eye by the interposition of screens (S). Fig. 1 shows the test set-up. By proper adustment of the screens we can stimulate "binasally" (each eye sees homonymous objects) or "bitemporally" (each eye sees heteronymous objects). Both methods of stimulation are shown in Fig. 2. The interval between stimuli was made equal to zero as near as possibile. When, under these circumstances, we apply alternate haploscopic stimulation to the eyes with a low frequency of alternation (*e.g.*, each retina receives 20 stimuli in 30 seconds), maximal apparent movement results. When the frequency of stimulation was raised we noticed the following phenomena:

The amplitude of the apparent movement was reduced, whereas the movement retained its optimal character; the subject still had the impression of one object moving over a certain distance. The distance across which the movement was seen, however, lessened.

A further increase of the frequency reduced the amplitude still more, until one object exhibiting slight movements was seen. Still further increase of frequency caused the movement to disappear completely and only one motionless object was seen. These effects resulted from a fusional movement; the motor impulses roused by the alternating retinal stimulation found a concrete expression in this fusional movement.

Increase of frequency, however, may also be followed by another phenomenon. Instead of apparent movement over a smaller distance, simultaneous perception of two objects at the original distance may result (*sim-stadium* of Wertheimer). This simultaneous perception of two objects is still accompanied by some apparent movement. The two objects appear to "jump" towards their place and away from it. ("Aufauch" and "Erlösch-Bewegung," Roelofs and Van der Waals.) A further increase of frequency then results in simultaneous perception of two objects separated in space, without any apparent movement.

Summarizing:

Low frequency—maximal apparent movement of one object.

Increase of frequency—either apparent movement over a smaller distance or simultaneous perception of two objects accompanied by "jump" apparent movement.

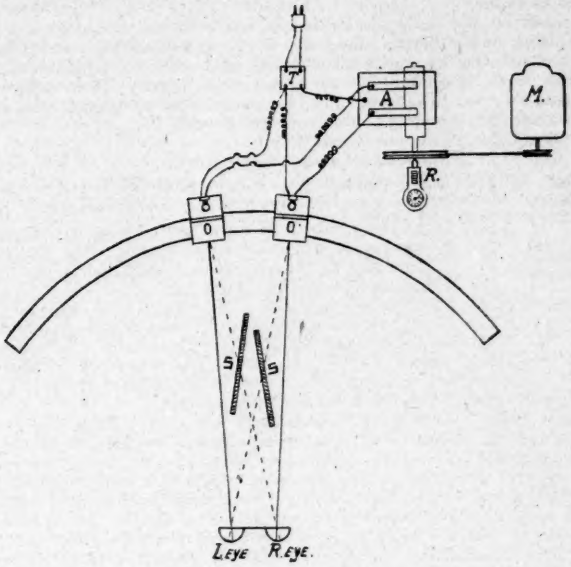

FIG. 1.

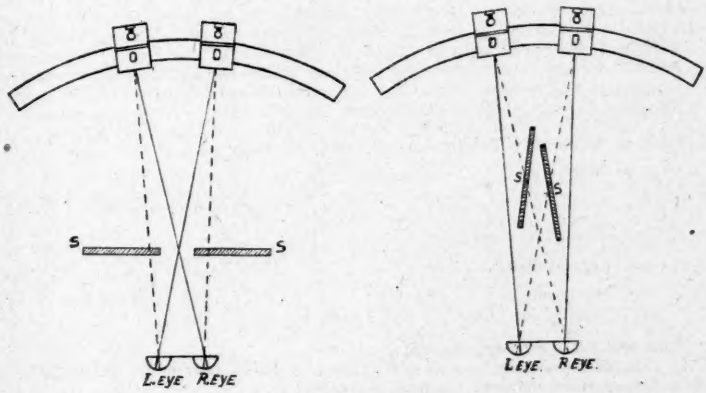

FIG. 2.

Further increase—either one motionless object or two motionless objects separated in space.

It depends on the outer circumstances which of the two reactions is chosen. The frequency of alternating stimulation is of great significance with regard to the reaction. The maximal apparent movement seen with low frequency is gradually transferred to the standstill of one object with increase of frequency. In our experiments we tried to establish the frequency of the alternating stimuli required for the different phases of the above mentioned process.

The following steps were paid attention to:

Increase of frequency
1. Maximal apparent movement (m)
2. Initial shortening of amplitude of the apparent movement—first symptom of a fusional movement—(m + f)
3. Complete standstill of the object—fusion is complete—(f)

Lowering of frequency
4. Reappearance of apparent movement after the standstill—described by the subject as "slight movement"—(f + m)
5. Maximal apparent movement (m)

The experiments were carried out with "bitemporal" as well as "binasal" stimulation (see Fig. 2). The distance between the subject and stimulating objects was 2,89 M. The distance between the objects corresponded to a visual angle of 1° 5′ (55 mm.) and also 2° 22′ 40″ (12 cm.). The frequency of stimulation is expressed in the number of stimuli per 30 sec. falling on each retina. The experiments were also carried out with a red and a green object.

In Table I the results of the experiments are given. Each line in the table contains the mean values of a series of experiments carried out in one session.[3] The apparatus was only used in preliminary investigations. In further experiments the objects were projected on to a screen by means of a projection-tachystoscope. In this method the wooden boxes with a 3 mm. diameter hole were substituted by luminous spots of 3 mm. diameter projected on to a screen. In all other respects the test set-up was the same as in the preliminary experiments. All figures of Table I were obtained by experiments with the projection-tachystoscope. Our subject had a slight exophoria (Maddox right eye: 36′; left eye: 24′).

	m+f	f	f+m	m
BITEMPORAL STIMULATION				
Visual angle 1°5′	85	114	103	27
	74	118	111	65
	75	117	100	58
	92	124	100	56
	96	126	108	64
Visual angle 2°22′40″	44	112	—	—
Coloured objects (red and green)	80	112	—	—
BINASAL STIMULATION				
Visual angle 1°5′	51	117	91	31
	58	120	99	26
	42	122	111	18
	32	112	—	—
	44	120	110	28
Visual angle 2°22′40″	21	112	—	—
	30	110	106	20

TABLE I

From this table we may conclude:

1. Alternate stimulation with a frequency of 10-20 stimuli per 30 sec. on each retina brings about maximal apparent movement.

2. A shortening of the amplitude of this apparent movement occurred on bitemporal stimulation with 74-96 stimuli per 30 sec. on each retina with objects subtending 1° 5′; for a visual angle of 2° 22′ 40″ a shortening occurred with 44 stimuli. On binasal stimulation these values were 32-58 stimuli/30 sec. for a visual angle of 1° 5′ and 21-30 st./30 sec. for a visual angle of 2° 22′ 40″.

These values mark the beginning of fusional movements.

3. One motionless object was seen when the frequency was 112-126 for bitemporal stimulation; and 110-122 for binasal stimulation.

These values marked complete fusion of the objects.

4. Lowering of the frequency made the movement appear again. Initial movement occurred with 91-111 stimuli on binasal stimulation and 100-108 stimuli on bitemporal stimulation. With this frequency the motor impulses were no longer strong enough to maintain complete fusion.

5. A further lowering of the frequency results in apparent movement with maximum amplitude. This occurred on bitemporal stimulation with a frequency between 17-65, and on binasal stimulation between 18-31.

6. With coloured objects (one eye was stimulated by a red, the other by a green object), a blending of colours occurred with the same frequency, which caused a fusion of the white objects. This suggested that motor and sensory fusion here occurred with the same frequency.

Fusional movement, however, did not always occur according to this scheme. Fusional movement was impaired especially when the darkness in the room was not complete. In these circumstances increase of frequency resulted in simultaneous perception of both objects, accompanied or not by "jump" apparent movement (see also page 630). In this case the motor impulses elicited by haploscopic alternate stimuli of both retinae followed a different reflex-path. The motor impulses did not bring about a fusional movement (reaction II, see page 630), but led to a localisation of both objects separated in space (reaction III, see page 630). The same phenomenon could be observed if one retina only were stimulated alternately by two objects. With low frequency, maximal apparent movement was seen, with increase in speed of alternation, "jump" apparent movement together with simultaneous perception of both objects separated in space resulted. A further increase gave simultaneous perception of two objects without any apparent movement. In case of stimulation of one retina, reaction III only is elicited.

We then investigated whether the same frequency, which brings about a fusional movement, may also by following another reflex-path give rise to the simultaneous perception of two objects separated in space. For this purpose we drew a comparison between haploscopic alternate stimulation of both retinae and alternate stimulation of one retina only. In case of the former the motor impulses follow the reflex-path of reaction II; with the latter it was reaction III.

We therefore investigated the effect of monocular alternating stimulation in the same way as in the binocular experiments. The right and left eyes were investigated separately and the investigations were performed with and without a fixation-point. The fixation-point made it possible to investigate the effect on the temporal and nasal half of the retinas and also the effect of one stimulus falling on the nasal half and the other on the temporal half of the retina. The objects subtended a visual angle of 1° 5′. The test set-up was the same as in the binocular experiments, the only difference being that one eye was covered and the screens were removed. The results given in Table II are quoted under five headings:

	m	m+s	s	s+m	m
WITHOUT FIXATION POINT					
R. eye	20	32	108	92	48
L. eye	24	44	92	80	40
WITH FIXATION—POINT					
nasal half of retina:					
R. eye	—	24	104	76	28
L. eye	20	48	100	96	16
temporal half of retina:					
R. eye	28	44	96	92	40
L. eye	—	20	120	96	16
temp. + nas. halves of retina:					
R. eye	20	36	88	72	32
L. eye	20	40	84	60	32

TABLE II

maximal apparent movement (m); increase of frequency: apparent movement + simultaneous vision (m + s); simultaneous vision without movement (s); lowering of frequency: simultaneous vision + apparent movement (s + m); maximal apparent movement (m).

These results were compared with the values of the binocular experiments quoted in Table I:

Monocular altern. stim.	Binoc. haplosc. binasal stim.	altern. stim. bitemp. stim.
m+s: 20-48	m+f. 32-51	74-96
s: 84-120	f: 110-122	112-126
s+m: 60-96	f+m: 91-111	103-111
m: 16 48	m: 18-31	27-65

Considering that it is very difficult for the subject to indicate when he sees two objects without apparent movement (s) and two objects with slight apparent movement (s + m), we may conclude that the values in the two columns fairly agree. These values indicate that the beginning of a fusional movement (m + f) and the beginning of simultaneous perception (m + s) occurred with about the same frequency of alternation. We may substitute fusion (reaction II) for simultaneous perception (reaction III). In both instances the stimuli are the same; they give rise to identical motor impulses which, however, may follow two reflex-paths—one leading to reaction II (fusion) and the other to reaction III (simultaneous perception). In case of monocular alternate stimuli only reaction III is available, but in binocular haploscopic stimulation reaction II or reaction III may be chosen. It depends on the outer additional circumstances which reflex-pathway is used. When the room was not completely darkened, reaction III (simultaneous perception of two objects) was favoured. In a following article we will prove that this was caused by additional peripheral stimuli from the surroundings. If the influence of these peripheral stimuli was excluded by using a completely darkened room, reaction II (fusion) always occurred.

These experiments concerned fusion and movement in the horizontal direction. We repeated the same experiments with the stimuli placed vertically, one above the other. The experiments for the vertical direction yielded results which were similar to those in the horizontal direction. To obtain fusion in the vertical direction, however, it is necessary to use a smaller distance between the stimulating objects.

Conclusion and Summary

A theory on the binocular optical reflexes is presented. Each retinal stimulus gives rise to a sensory *and* a motor impulse. This motor impulse may be responsible for the following reactions:

(I) A sidewards movement of both eyes in the same direction.

(II) A movement of both eyes in opposite directions. These movements may, however, be inhibited, in which case the motor impulse gives rise to

(III) Localisation of the stimuli separated in space,

(IV) The perception of apparent movement.

An investigation of reactions II, III and IV was carried out by means of alternate haploscopic stimulation of both retinae. By varying the speed of alternation of the stimuli, it was possible

to analyze the conditions for fusional movements. Haploscopic stimulation with low frequency of alternate stimuli gives rise to reaction IV (maximal apparent movement). Increase of frequency results in a combination of reaction IV with reaction II (apparent movement combined with a fusional movement) or in a combination of reaction IV with reaction III (apparent movement together with simultaneous perception). A further increase of frequency causes reaction IV to disappear and fusion (reaction II) or simultaneous perception of two objects (reaction III) results. Reactions II and III occur with the same frequency of alternate stimuli and are brought about by motor impulses of the same quality and quantity. These motor impulses may follow different reflex-paths leading to reaction II or reaction III. Additional circumstances may inhibit one reflex-path and favour the other; *i.e.*, alternate haploscopic stimulation of both retinae in a completely darkened room results in reaction II; if the room is not completely darkened reaction III is favoured.

According to this theory and based on experimental evidence, fusion, localisation and apparent movement are explained as physiological rather than psychological phenomena. They are optical reflexes, of which the physiological correlate is provided by the motor impulses elicited by the retinal stimuli.

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HYALINE MEMBRANES ON THE POSTERIOR CORNEAL SURFACE

BY

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LONDON

HYALINE membranes attached to the posterior corneal surface and spanning the anterior chamber are striking curiosities. They may arise (1) as persistence of embryonic tissues, (2) as detachments of Descemet's membrane and endothelium, or (3) as sheets of inflammatory exudate; the following three cases illustrate each of these groups, and the aetiology is discussed.

CONGENITAL HYALINE MEMBRANE

Case History. A man, aged 31 years, whose right eye had been divergent and almost blind since infancy, showed the following abnormal signs in that eye.

Behind the axial area of the cornea lay a semilunar transparent membrane, joining the cornea along a line that passed from 11 to 7 o'clock, roughly concentric with the limbus, 1·3 mm. internally, and merging there with the posterior corneal band. The free margin ran vertically to connect either end of the attached margin: in the centre of this free margin, overlying the pupil, was a small area of pigmented tissue, resembling persistent pupillary membrane, and from it a brown cord passed deeply and downwards to reach the lesser circle of the iris at 6 o'clock. There were no keratic precipitates, and the corneal diameter was 12 mm., as compared with 13·5 mm. in the other eye. The pattern of the iris was less well marked than in the other eye, and there were three clumps of axial epicapsular stars on the lens. The eye was highly myopic (−9·0 D.), and even this correction did not improve his vision beyond finger-counting. There was a divergent concomitant strabismus of 20°, rectified by operation, and a coarse latent nystagmus.

His father and grandmother also had divergent squints, but the father's eyes were otherwise normal.

Abscission of the membrane was performed on July 7, 1948; the attached edge was divided by the sweep of a Ziegler's needle-knife, and a few days later the whole membrane was lifted out through a keratome section, the cord to the iris being divided with scissors. It now became possible to see the fundus, which showed extensive myopic choroido-retinal degeneration, and no visual improvement ensued. (See Figs. 1 and 2.)

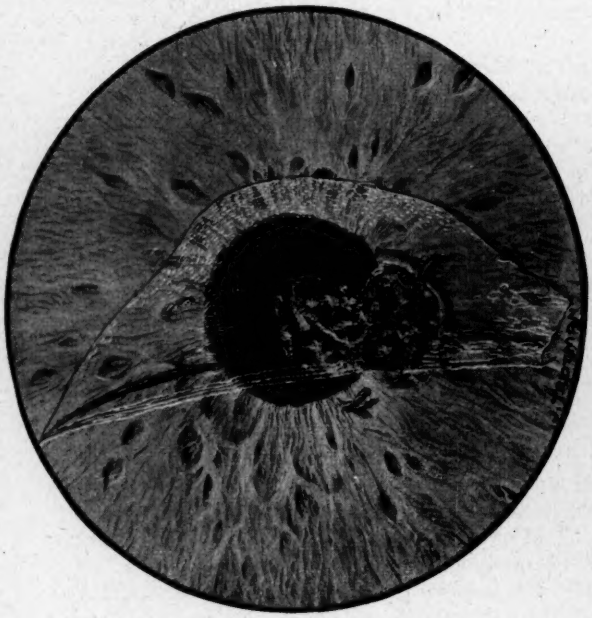

FIG. 1.

FIG. 2.

DISCUSSION. During development the lens vesicle becomes separated from its parent surface epithelium, and the intervening space is traversed by the protoplasmic threads of the mesostroma or the anterior vitreous body, which is ectodermal in origin. This mesostroma is most evident peripherally, filling the space where the curve of the lens bends away from the surface ectoderm. A directional membrane is formed by a condensation of the mesostroma, whose anterior surface remains in continuity with the epithelium, but whose posterior surface becomes separated from the lens by a space created by atrophy of the cones of the lens epithelium, from which it partly originated. The mesodermal masses lie peripherally, and grow in centripetally both along the posterior surface of the directional membrane as the future endothelium of the cornea; and later, anteriorly to the endothelium, appearing as a wedge of nuclei separating endothelium from epithelium, and ultimately compressing the mesostroma, which it invades, into the condensations of Bowman's and Descemet's membranes. Finally the mesoderm of the iris and pupillary membrane grow in from the post-endothelial mesodermal condensations.

Persistence of the post-endothelial tissues may occur, and thus present as a fluffy mesh occluding the angle and liable to produce glaucoma; the congenital hyaline membranes described in the literature—Mann (3 cases), Ballantyne, Clapp, Hagedoorn —are all of this order, and in all of them the hyaline membranes were present beneath the cornea at its periphery only, and never extended more than a few mm. from the limbus. Only Ballantyne's case (six days old, with lid and iris colobomata), had keratic precipitates. The other patients were more mature, three showing characteristic thickened ridges on the iris, and one a band which passed forwards to the cornea. The post-endothelial tissue is abundant only peripherally, where these anomalies developed, but the foregoing case, where only central anomalies are present and the characteristic iris anomalies are absent, is not easy to explain in this way.

The mesostroma over the centre of the lens is very thin, as the lens remains almost in contact there with the parent ectoderm, separated from it only by the endothelium, and consequently an incomplete separation of the membranous pupillary membrane from the endothelium is likely to occur, with the result that an anterior synechia of pupillary membrane to endothelium, and subsequently of the mesodermal pupillary membrane and secondarily of the iris will result. This would seem to explain the origin of the case I have reported; for it may be surmised that subsequent development of the eye, with deepening of the anterior

chamber, caused the endothelium and Descemet's membrane to be thereby stripped off the cornea by traction of the synechia over the area where the anterior chamber was deepest.

The cornea of the affected eye was significantly smaller than its fellow, in spite of the high myopia, and a similar feature was noted in one of Miss Mann's patients. If it had been unduly large, one might have guessed that the membrane was stripped off like a retinal detachment in an elongating myopic eye, but the small cornea is presumably evidence only of underdevelopment of the anterior segment, as often happens with persistent embryonic tissues. Such congenital membranes might result from an intra-uterine inflammation, with the anterior iris synechia as evidence of this, and the membrane as a secondary detachment through traction; but apart from the synechia (which in both the foregoing case and the case of Miss Mann's in which it was described was large, single, and free from inflammatory irregularities) and the keratic precipitates in the infant described by Ballantyne (which does not fall so readily into this group), there were no signs of inflammation. The membrane reported bore little resemblance to the true inflammatory sheets of interstitial keratitis, etc., as will be evident when this group is discussed.

Divergence of the eye in the above case is probably not due to impaired binocular vision caused by the membrane, but coincidentally to an hereditary monocular myopia, since his father and grandmother had divergent eyes but no membrane.

Detachments of Descemet's Membrane

Hyaline membranes can be present since birth, not as a congenital defect, but resulting from birth trauma after distortion of the eye with forceps: into this category falls the following case, where a very similar membrane was found behind the centre of the cornea in an eye which had also been divergent and weak-sighted since infancy.

Case History. A man, aged 20 years, with a divergent and partially blind left eye, ascribed his disability to a forceps delivery.

A semilunar transparent membrane lay behind the central area of the cornea, joining it along a line that passed from 10.30 to 5.30 o'clock from the corneoscleral margin at either end, but 3 mm. internal to it laterally; there it merged with the posterior corneal band. The free margin connected the ends of the attached margin, and was curled over on itself, so that it appeared double in the slit-lamp beam. Another line curved nasally from the upper limit of the membrane to just short of the lower limit, and appeared to be a thickening of Descemet, skirting peripherally a similar semilunar area. A dust-like opacity was present in the middle of the corneal thickness over the whole pupillary area; there were no keratic precipitates. A pulverulent opacity was present in the anterior foetal nucleus of the lens. The iris and fundus were normal. The eye

was highly astigmatic (−1·5 D.S., +5·0 D.C. at 110°), but this correction did not improve his vision beyond finger-counting.

A 40° divergent concomitant squint was rectified by operation. The right eye was normal and emmetropic, and there was no relevant family history. (See Figs. 3, 4 and 5.)

FIG. 3.

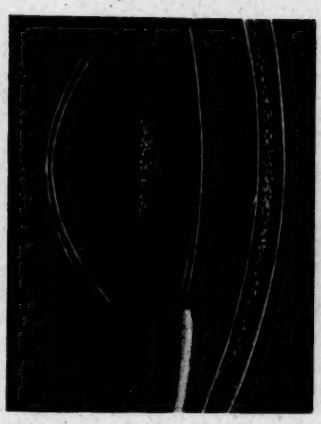

FIG. 4.

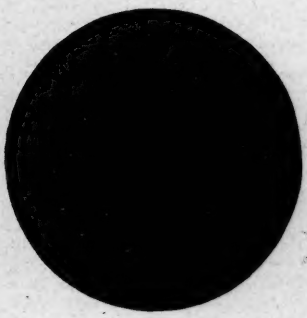

FIG. 5.

DISCUSSION. Thomson and Buchanan described three varieties of birth injury to the cornea, in all cases following difficult labour and the use of forceps: (1) A diffuse opacity which is temporary, and due to oedema. (2) A similar diffuse opacity which becomes permanent through superadded inflammation. (3) An opacity which is linear and permanent, and due to rupture of Descemet with or without the posterior corneal lamellae. Subsequently cases were recorded in which tears of Descemet were followed by its detachment over a wide area. This usually produced a series of vertical ribbons between the parallel tears, which were attached only at their upper and lower extremities (Peters, Feingold, Lloyd, Rushton, Perera, Fison): these bands tended to roll up into rods if they were narrow, so that a colonnade seemed to grace the forecourt of the eye. In other cases, as in that described here, a falciform or segmental detachment was present, again with the long axis roughly vertical. (Ballantyne, Fewell.)

All such birth injuries of the cornea are becoming less common as rickety pelves become rarer, and accoucheurs become more deft and less reliant on forceps. These lesions occur predominantly in the left eye, as in the above case and in Lloyd's five cases, since the left occipito-anterior position is commonest, bringing the left eye laterally. The damage is probably caused by pressure directly on the cornea by the long axis of the forceps blade—so everting and vertically splitting the inner layers.

In the above case the nasal edge of the torn Descemet's membrane has remained (as usual) as a small ridge through the greater part of the extent of the original tear, while temporally the membrane has become detached over a wide area. The fine powdery opacity in the *substantia propria* of the area that has been denuded is typical of those cases of more severe damage, where some fibrosis has followed the initial oedema. There is also the characteristic high astigmatic error (5·0 D.C.) in the slightly oblique axis of the tear, such uni-ocular astigmatism being sometimes attributed *per se* to obstetric pressure; and there is characteristically no mesodermal abnormality.

INFLAMMATORY RETROCORNEAL MEMBRANE

Case History. A man, aged 59 years, with a classical history of interstitial keratitis—bilateral "blindness" at the age of 13 years, which after two years cleared gradually in the left eye only, and an acknowledged parental infection with syphilis—showed the following appearance.

The right eye was blind, and directed 15° upwards and 15° convergent. Both corneae were slightly ectatic, with characteristic interstitial nebulae, some deep attenuated blood-vessels, and, in each eye, an eccentric deposit of blood pigment. Over the back of the right cornea stretched a reticulate sheet, the interlacing fibrinous bands being as wide as the holes that separated them. They adhered to the posterior corneal surface except at a very few points, where a gap between

band and Descemet's membrane was discernible; these bands became fainter peripherally, where the cornea was relatively clear.

Over the back of the left cornea were fewer but more discrete interlacing strands, spanning the anterior chamber like a cobweb so that centrally they lay well behind the cornea. Inserted, as they were, into the latter at either end, they seemed to consist of a fine opaque white central filament with a relatively broad translucent sheath.

Both irides were atrophic, especially on the right side, with posterior synechiae also on that side. Exudate on the anterior lens capsule, cortical cataract and a small bound-down pupil prevented a view of the right fundus, and this eye had no perception of light. The left lens was clear except for a few epicapsular stars. The left fundus was normal centrally, but showed extensive peripheral disturbance from old anterior choroiditis. The left vision was improved to 6/18 with −5·5 D.S. correction.

In spite of having had no specific treatment, he was otherwise healthy, except for deafness on one side. His Wassermann reaction was negative. His mother and father had "died of drink" when he was young, but his brother, aged 66 years, was alive and well, and also showed on examination a typical interstitial keratitis, although much less intense, the vision being 6/60 and 6/24 with myopic correction. There were no comparable abnormalities in the brother's eyes, except for a fine translucent strand passing in the right eye across the pupil from one posterior synechia to another, and jutting forward into the anterior chamber, so that it was well clear of the lens capsule. (See Figs. 6, 7, 8, 9.)

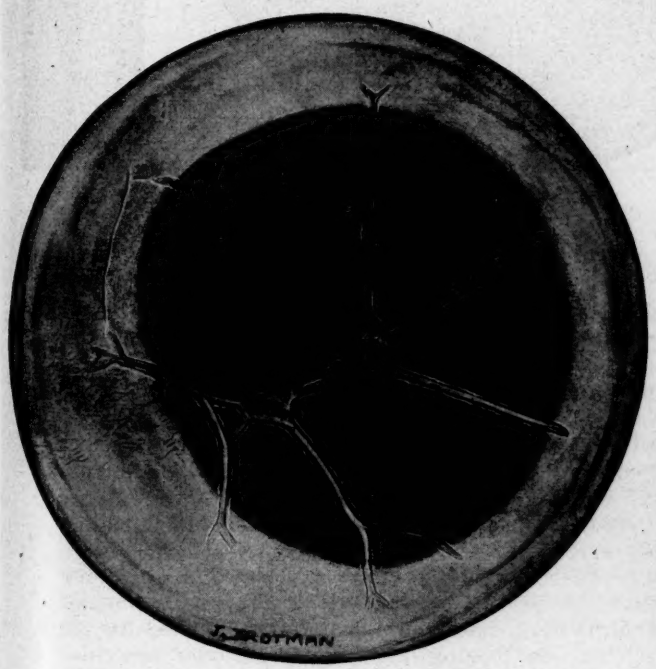

FIG. 6.

FIG. 7.

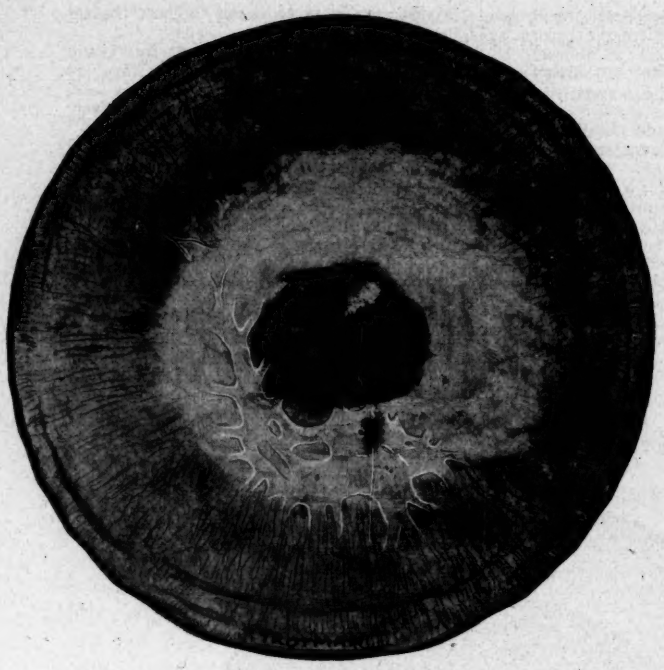

Fig. 8.

FIG. 9.

DISCUSSION. Membranes from inflammatory exudate are considerably less rare than the two varieties already described, and are more frequently recognised in the literature, so that little comment is here required. Typically they are seen as irregular fibrinous laminae, reticulate or stellate, and not as an even and thin semilunar sheet. Commonly they admit the term "glass membrane."

They occur classically in interstitial keratitis, but are occasionally described in tuberculous or other forms of intense uveokeratitis. They lie behind the centre of the cornea, and are necessarily associated with other signs of past inflammation. Initially they were probably wholly in contact with the endothelium, but become detached like the strings of a bow when their fibrin content subsequently retracts. It is of passing interest to note the fibrinous filament in the anterior chamber of the brother's eye, but here passing between two posterior synechiae instead of between two points on the posterior corneal surface.

Perhaps the precipitation of fibrinous shreds depended on an hereditary predisposition or on the particular strain of spirochaete.

My acknowledgments are due to I. C. Mann for necessary advice and assistance, and to E. F. King and A. G. Cross for permission to use their case-records.

Summary

Three cases are described of hyaline membranes; one of them a consequence of developmental arrest, one due to birth trauma and the third following uveokeratitis. Their aetiology is discussed.

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LOCAL APPLICATION OF UREA FOR THE TREATMENT OF DENDRITIC ULCER*

BY

M. KLEIN *and* S. J. H. MILLER

LONDON

HYPERTONIC solutions applied in an eye-bath are believed to have a favourable influence on marginal ulcers of the cornea. One obvious advantage of an eye-bath is that the medication is kept in the conjunctival sac. A series of cases of dendritic ulcer was treated with a corneal bath containing a 25 per cent. solution of urea in 1942-44 at the Central London Ophthalmic Hospital and the results in over 30 cases were encouraging.† The same treatment has been carried out in a new series, and the results are reported here. It

* Received for publication, April 11, 1949.

† M. Klein. Results unpublished, as the notes of these cases were lost during the war.

appears that urea, though widely used in other branches of medicine, has not been employed in ophthalmology.

Historical.—The effect of urea on bacteria has been known for many years. Ramsden (1902) noted that urea inhibited putrefaction. According to Peju and Rajat (1906) urea added to a nutritive medium produces abnormal morphological forms, and if the concentration is increased the growth of bacteria stops altogether. Wilson (1906) noted that 1·5 to 3·5 per cent. of urea in the medium caused pleomorphism, and an addition of 8 per cent. urea prevented the growth of B. coli. In 1915 Syms and Kirk observed that 25 per cent. or stronger solutions of urea killed B. typhosus, and local applications of crystalline urea resulted in the sterilization of wounds. Foulger and Foshay (1935) treated purulent otitis media by local application of saturated solutions of urea, and McKay and Schröder (1936) found that concentrated urea solutions inactivated rabies and poliomyelitis virus in a relatively short time. The use of urea gained popularity recently when it was found that it increased the activity of locally applied sulphonamides, and a dusting powder consisting of urea and sulphonamide was widely used for wound treatment during the late war. Weinstein and his co-workers (1944-1947) studied the antibiotic activity of urea and its related compounds (urethane and thiourea) and found that the antibiotic effect of these preparations is considerable and is mainly directed against Gram-negative bacteria. Gram-positive bacteria are affected only by exposure to higher concentrations and for longer periods (Weinstein and co-workers). Some of the urea derivatives, especially phenylurethane, can be regarded as poisons acting directly on the cell. Spores soaked in 0·5 per cent. urethane show effects similar to irradiation with 1,000 r X-rays (Haddow 1948). Urethane has also been administered in leucaemia and the effect is said to be comparable to that of deep X-ray therapy. (Paterson *et al.*, 1946.)

Properties of urea and its derivatives are as follows: Urea forms white hygroscopic crystals. Solutions must be kept sterile as they are liable to undergo ammoniacal decomposition if contaminated. Urea is a normal constituent of tissues and blood. It is not toxic, and if it is applied to wounds, healing is not interfered with. Thiersch grafts do not necrose in the presence of urea. Urea solutions are neutral and their antibiotic activity is retained in acid or alkaline media. Urea penetrates cells readily, *e.g.*, red blood cells, and the urea content becomes similar within and without. It does not cause haemolysis. The break-down products of tissues are dissolved in a solution of urea, a point of importance in the local use of sulphonamides. Clinically urea

solutions have proved useful in chronic purulent wounds; they stimulate healing and help in the separation of sloughs (Williams, 1946 and Robinson, 1936). Urea can be used with advantage in combination with sulphonamides, as it enhances their solubility and contributes its own bactericidal effect. It can also be used in combination with penicillin, and, according to Weinstein, urea limits the production of penicillinase. Derivatives of urea which have been used or investigated for antibiotic effects are : allantoin, urethane and phenylthiourea. The action of urea on wounds is the rationale of the present study.

LOCAL USE OF UREA IN CASES OF DENDRITIC ULCER

As in the preliminary series, the following method of treatment was used in the present series. The eye was anaesthetised with two instillations of 0·5 per cent. amethocaine hydrochloride B.P.

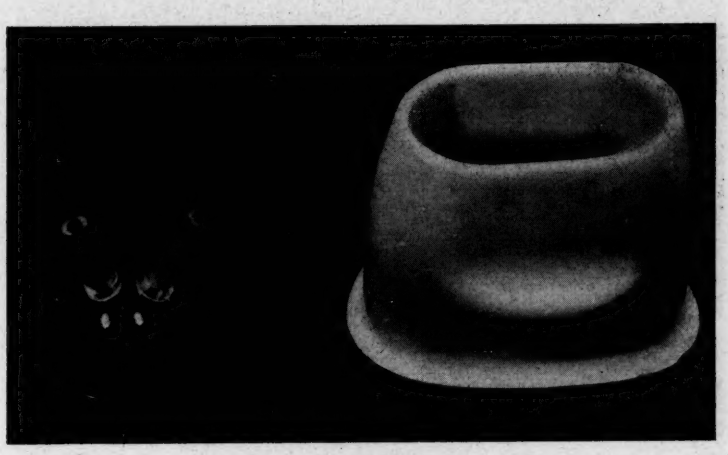

FIG. 1.

(*a*) Contact shell applicator. (*b*) Eyebath.

The contact lens applicator* was placed over the eye (Fig. 1a and Fig. 2) and filled with a freshly made solution of 25 per cent. urea to which 2 drops of amethocaine was added to each 5 ml. solution. The application was well tolerated and treatment was done either in the sitting or recumbent position. Through the transparent plastic contact applicator the eye was watched during treatment, and if the ulcer was not covered by the fluid the appli-

* Made by G. Nissel & Co., Ltd.

cator was replenished. The duration of the treatment was 5 minutes. Immediately after treatment the cornea usually showed diffuse staining round the dendritic ulcer, and very often a large adjacent area became denuded of epithelium.

In some cases there was difficulty in the use of the contact applicator, and a bakelite eyecup, the bottom of which was removed, was used instead (Fig. 1b and Fig. 3). After placing the cup over the open eyelids, the urea solution was poured into the cup. By

FIG. 2.

Contact shell applicator inserted without fluid. Condensation shows up the raised corneal portion.

a slight pressure with the edge of the cup the palpebral fissure was kept open so that the cornea was bathed with the fluid. With this treatment the patient had to be in a recumbent position.

The urea treatment was followed by instillation of 1 per cent. atropine drops, yellow soft paraffin was placed in the lower fornix and the eye bandaged. The dressing was changed after 24 hours, when the staining area was found to have diminished or completely healed over. A second or third urea treatment could be given with an interval of 24 hours between each treatment. A mild ointment such as yellow soft paraffin was useful in preventing the breaking-down of the newly formed epithelium.

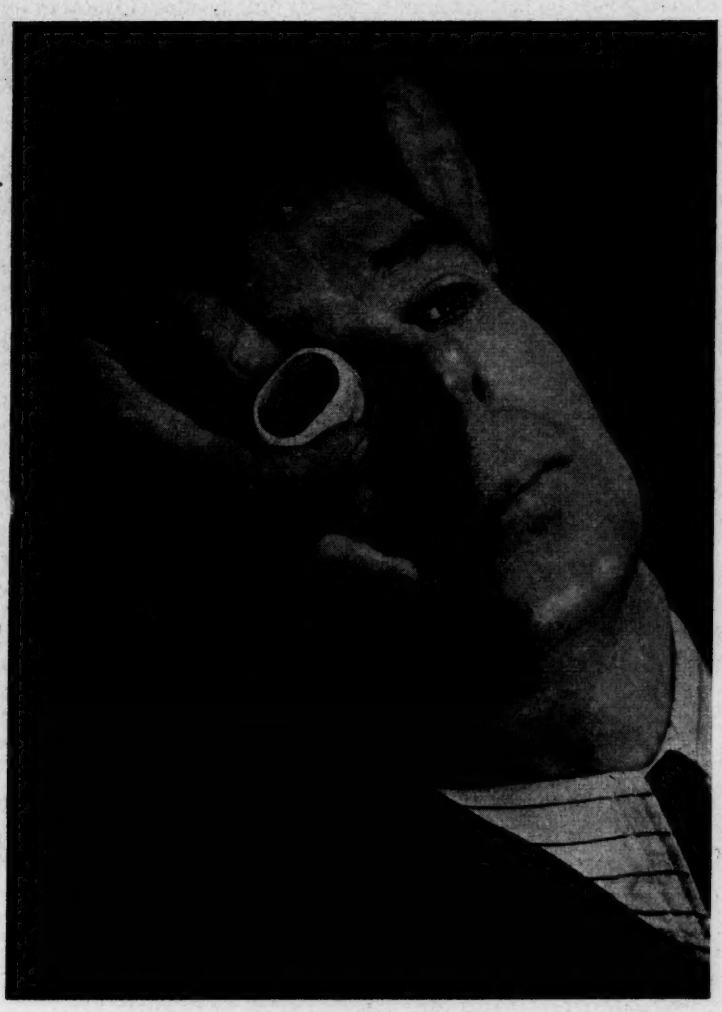

FIG. 3.

Eyebath in use. The edges of the cup hold the palpebral fissure open.

RESULTS

Thirty-three cases were treated in this series arbitrarily divided into three groups according to the severity of the lesion. Group I included all the cases in which there was a well localized dendritic

TABLE I

Data on individual cases

No.	Age	Sex		Severity			No. of urea applications	Healing time in days	
		M.	F.	I	II	III			
1	45	M.	—	—	II	—	1	13	—
2	70	M.	—	—	—	III	—	40	See Remarks
3	30	M.	—	—	II	—	2	14	—
4	28	M.	—	—	—	III	2	28	See Remarks
5	35	—	F.	—	II	—	2	42	Healed 12 days after urea, See Remarks
6	71	M.	—	I	—	—	1	2	—
7	58	M.	—	I	—	—	1	2	—
8	46	M.	—	I	—	—	1	4	—
9	67	M.	—	I	—	—	1	3	See Remarks. Urea treatment was given 4 weeks after onset and 3 weeks after other treatment had failed.
10	62	M.	—	I	—	—	1	10	History of previous dendritic ulcers 1933, 1943, 1946.
11	63	M.	—	I	—	—	1	5	—
12	60	M.	—	—	II	—	1	28	History of previous dendr. ulc. See Remarks.
13	25	M.	—	—	II	—	1	7	Came to Hospital 1 month after onset.
14	62	M.	—	—	II	—	1	9	History of previous dendritic ulcer.
15	19	—	F.	I	—	—	1	7	—
16	38	M.	—	I	—	—	1	7	—
17	48	—	F.	—	II	—	1	28	Transient punctate staining of cornea. Hist. of prev. dendritic ulcer.
18	45	—	F.	I	—	—	2	16	Transient punctate staining of cornea.
19	61	M.	—	I	—	—	1	15	Transient punctate staining of cornea History of prev. corneal ulcers, ? nature.
20	24	M.	—	I	—	—	2	5	—
21	51	M.	—	I	—	—	1	7	—
22	51	M.	—	I	—	—	1	7	—
23	39	M	—	I	—	—	1	10	—
24	64	M.	—	I	—	—	1	11	—
25	70	M	—	I	—	—	1	5	—
26	68	M.	—	—	—	III	1	42	See Remarks
27	69	M.	—	—	II	—	1	13	Transient punctate staining.
28	52	—	F.	I	—	—	1	—	No response to urea, transferred to Radiotherapy
29	38	—	F.	I	—	—	1	7	—
30	38	—	F.	I	—	—	1	9	—
31	44	M.	—	I	—	—	1	17	—
32	53	M.	—	I	—	—	1	13	—
33	56	M.	—	I	—	—	1	6	—

figure, Group II those in whom the lesion was more extensive, and Group III those with deeper lesions marked either by infiltrations of the deeper layers of the cornea, or by cells in the anterior chamber, or by hyperaemia and swelling of the iris without patent herpetic iritis.

REMARKS

(2) Recurrent dendritic ulcer; attacks almost every year since 1933. No response to treatment. Iodised six times, carbolised twice, urea applied three times, vaccinated, sulphacetamide locally applied, retrobulbar alcohol injected. Eventually the eye became quiet after tarsorrhaphy. Duration 40 days. Urea ineffective.

(4) First urea treatment on the fourth day after onset. The eye made a slow recovery and on the 21st day the ulcer recurred, for which a second treatment with urea was given. Two days later there was no staining. Received two urea treatments; healed in 28 days.

(5) October 17, 1947. Carbolised and local penicillin applied: October 20, 1947, vaccinated. One month after treatment had failed urea was applied, and 10 days later repeated. Rapid healing followed in a few days. A fortnight later a small staining area was noted, urea treatment was given again and healing followed.

(9) Iodised twice, local albucid applied, without success. Three weeks after treatment had failed, urea was applied and 3 days later there was no staining. Four weeks after discharge there was a relapse which healed without treatment in one week.

(12) After the first urea treatment the condition improved but the epithelium was constantly breaking down. Patient was treated with albucid, healing was slow, taking 4 weeks. History of dendritic ulcers previously.

(26) Had one urea treatment. After some improvement, the ulcer broke down and did not heal until intravenous ascorbic acid (500 mg. daily for 7 days) was given.

Group I was the largest with 22 cases. The number of days needed for healing varied between 2 and 16 after the application of urea, with an average of 8 days. In 20 cases one application of urea proved sufficient. The average healing period of 8 days applies to cases in which 6 or 8 days after the urea treatment, punctate staining appeared. In these cases the time was recorded when the corneal condition was completely healed.

Group II contained 8 cases. Healing time was protracted in two cases owing to a recurrence of the staining area. In the remaining cases of this group the healing time was from 7 to 14 days with an average of 15·5 days for the group as a whole.

TABLE II

Classified data on the patients in group

	Healing time in days			Distribution of age			Total	Distribution of sex	
	average	min.	max.	under 35	36-55	56 & over		Male	Female
Group I	8	4	16	2	11	9	22	18	4
Group II	15.5	7	42	3	2	3	8	6	2
Group III	36	28	42	1	—	2	3	3	—
				6	13	14	33	27	6

Three cases fell into Group III. The healing period was in one case 28 days, in the other two 6 weeks.

Discussion

In this series, dendritic ulcer was predominantly a disease of males, and mostly of the older age group, as can be seen from Table II. In those cases giving a history of one or more attacks of dendritic ulcer in the past the tendency to healing was poor. On the other hand, where the dendritic ulcer developed upon corneal scars of other than dendritic origin, the healing time was the same as on a previously healthy cornea. After urea treatment the epithelium quickly regenerated, and the ulcer healed, but there was a tendency to relapse, a well-known characteristic of dendritic ulcer. It was therefore deemed important to give protection to the corneal epithelium by placing in the lower fornix a mild lubricating ointment and to keep the eye bandaged for 6 to 10 days after complete epithelialization. Patients were then advised to continue the pad and bandage at nights only, for another 3 weeks.

An important factor in the healing of dendritic ulcers is the general condition of the patient; the most severe cases appeared to be in poor physical condition, and the corneal lesion healed only after general treatment was given. In two severe cases, 500 mg. ascorbic acid was given intravenously daily for 7 days, and rapid healing was observed. It appears that the essential points in the treatment are (1) the favourable effect of locally applied urea followed by (2) protection of the healed area by a lubricant and a bandage, and in severe cases by (3) general treatment in order to improve the physical condition of the patient.

The punctate staining which was seen in a few cases following urea treatment may have been caused by the use of urea from a stock solution; it is advisable to use solutions freshly made. Recently in the treatment room, packets of 1 gm. urea powder have been kept ready, and the solution made by adding 4 ml. distilled water immediately before use.

Summary

The use of 25 per cent. urea solution applied as a corneal bath by means of a contact applicator or an eyecup is suggested as a method of treating dendritic ulcer. This application lasts from 5 to 10 minutes. The urea solution should be freshly made. The urea treatment is combined with treatment by atropine, yellow soft paraffin and bandaging of the eye.

No damage was observed from the application of urea. Results of 33 cases are reviewed. The milder cases generally healed after one application.

Our thanks are due to the Surgeons of Moorfields, Westminster and Central Eye Hospital, High Holborn, for permission to treat their cases. We are indebted to Mr. W. E. S. Bain and Mr. D. Langley for their help in the treatment and for observation of some of these cases, and to Sisters E. Kitchen and P. Hollis. For the photographs we are indebted to Mr. Peter Hansell.

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THE RELATIVE IMPORTANCE OF DIRECT AND INDIRECT OPHTHALMOSCOPIC EXAMINATION IN THE TREATMENT OF RETINAL DETACHMENT*

BY

H. ARRUGA

BARCELONA

IT is essential to employ both methods of ophthalmoscopy to obtain the highest possible percentage of successes in the surgical treatment of retinal detachment. I have seen on my travels that many colleagues employ one method to the exclusion of the other, and that in some countries the majority of the ophthalmologists use one method almost exclusively. If we grant the enormous importance of ophthalmoscopic examination in the treatment of retinal detachment, we must also make as complete an examination as possible by this method. I have seen colleagues who, though excellent clinicians and surgeons, only reached 55 per cent. of successes in these operations, owing, in my opinion, to the exclusive use of direct ophthalmoscopy as a mode of operative orientation.

These facts decided me to prepare the statistics of 200 unselected cases of my own, which I had classified in the following four groups:—

The *first* group includes those cases in which tears were visible to either method of examination. In the *second* group tears were visible only to indirect ophthalmoscopy, and in the *third* only visible

* Received for publication, May 27, 1949.

to direct ophthalmoscopy. The *fourth* group includes cases in which tears were invisible to both methods.

The figures are as follows:—

Cases with tears which were visible by both methods	154
Cases with tears which were visible only by indirect ophthalmoscopy	25
Cases with tears which were visible only by direct ophthalmoscopy	3
Cases with tears which were invisible to both methods	18
Total	200

Of the 18 cases forming the fourth group, 11 were extremely old ones, with opaque vitreous or other opacities, and 4 of them were clearly cases of exudative choroiditis.

The advantage of indirect ophthalmoscopy can be deduced from these statistics. Direct ophthalmoscopy is an excellent method for appreciating doubtful details, such as haemorrhage, and in order to measure differences of retinal level. Indirect ophthalmoscopy, however, with its larger field, better view of the periphery, and the possibility of employing a high light intensity of 150-200 candle power, is the method which should be preferred, although one should not use it exclusively. Pictures are reproduced of six cases in which it was possible to make a drawing of the retinal tear. In the remaining cases the tear was too obscure to be truly represented in any drawing without falsifying the reality, and these latter cases formed the majority.

EXAMPLES OF CASES WITH TEARS DIFFICULT TO DIAGNOSE

1. Tear near the ora serrata only visible with the indirect method. This represents a recurrence in an operated case. It was cured by diathermy.

2. Between the folds of the retina appeared a red line like a vessel. This sometimes disappeared, and it was only possible by the direct method to verify that it was a tear. During the operation the tear was half opened.

3. In the centre of the figure in a fold of the retina a tear with pigmented borders was recognised through the difference in level of its borders, no red coloration being seen. During the operation the tear was easily visible.

4. Detachment with large lateral disinsertion. The macula appeared perforated, but with direct ophthalmoscopy it was revealed as a cystoid degeneration without difference of level.

5. Tear only visible by indirect ophthalmoscopy, difficult to verify because there was a shallow detachment of the adjacent retina. The operative closure of the tear confirmed this diagnosis.

1
2
3
4
5
6

6. Tear near the ora serrata invisible with direct ophthalmoscopy. Except for the folds of the retina, it appeared like a spot of chorioretinitis. Closure of the hole cured the detachment.

Professor Dr. JOSEF MELLER (Vienna)

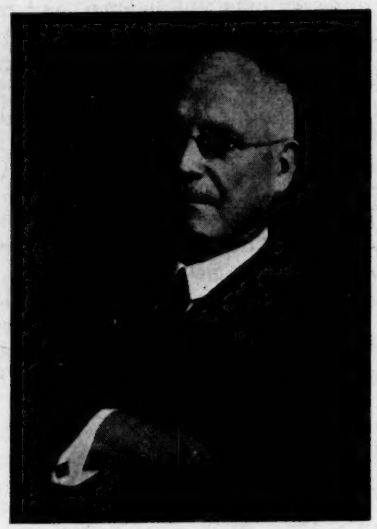
It is pleasant to salute outstanding men, and readers will have noticed that several recent articles in the Journal were dedicated to the subject of this address. On behalf of his British colleagues. we thank Professor Meller for the example he has set, and in offering him congratulations on his seventy-fifth birthday (October 22, 1949) we wish him pleasant days in retirement. His work and character will be remembered for many years. On the occasion of the seventieth birthday of Ernst Fuchs, Meller wrote about his predecessor : " His name shines forth in our science as that of his great teacher von Arlt." Meller in his turn has carried on the outstanding work of the Vienna School.

Since 1898 until his retirement in October, 1944, he worked at the Eye Clinic of Vienna, except for a three-year period (1915 to 1918), when he was " ordinary " professor of ophthalmology at the Imperial and Royal University of Innsbruck, to which he had been appointed by the Emperor Francis Joseph. In 1918 he was appointed " ordinary " professor of Ophthalmology at the Imperial and Royal University in Vienna, but by a letter from the German Reichminister for Science and Education, Berlin, dated May 10, 1944, he was relieved of the post of leader and teacher in the Vienna Eye Clinic " in the interest of later development at the high schools," and he retired from his position on October 1, 1944.

Unfortunately he seldom visited this country, although he had a good command of English, but he attended the 50th Annual Congress of the Ophthalmological Society of the United Kingdom at London in 1930 and became known to many of the participants who recollect his qualities of sincerity, modesty and geniality. He was also seen at the Oxford Congress a few times, and contributed to important discussions at the meetings of that body. Many will remember his delivery of the 1934 Doyne Memorial Lecture, which emphasised the continental view that iridocyclitis is often tuberculous in nature (as is now agreed in this country). He also reported that in some such cases tubercle bacilli may be recovered from the blood and from the eye, and argued that sympathetic ophthalmia is also probably tuberculous—dicta not accepted without considerable reserve.

Meller succeeded Ernst Fuchs at Vienna, and maintained the outstanding reputation for clinical work, research and teaching which that centre had deservedly earned. The professor and his assistants spoke English, and held post-graduate classes which were especially appreciated by students from the United States.

His fame brought honours to him, and he was an honorary member of the Academy of Science in Vienna. He was an



honorary member of the Ophthalmological Societies of the following countries: Hungary, Yugo-Slavia, Italy, Egypt, Brazil, Mexico and Greece. He was an honorary member of the Oxford Ophthalmological Congress and a recipient of the Doyne Medal. A list of his publications contains 124 items, mostly in the German literature between 1900 and 1941. To the English-speaking world he is best known by his book on ophthalmic surgery, of which the second edition was published in Philadelphia in 1912. It had an extensive sale for some years in this country. In Austria and Germany the " Augenärtzliche Eingriffe " ran to five editions.

F. A. J.

CORRESPONDENCE

THE FINAL RESULTS OF SQUINT OPERATIONS, IN WHICH RESTORATION OF BINOCULAR SINGLE VISION WAS NOT EXPECTED

To the Editorial Committee of

THE BRITISH JOURNAL OF OPHTHALMOLOGY

DEAR SIRS,—Dr. Stanworth is to be congratulated on this article which serves to emphasise the value of early and accurate operation in cases of strabismus in children.

He has not, however, perhaps sufficiently emphasised the importance of the age of onset of the condition. For instance, if a squint does not develop until after the age of three years, whatever the subsequent findings on the synoptophore, restoration of binocular single vision ought to be possible, provided that the visual acuity of each eye is approximately equal (or has been rendered so by means of occlusion) and provided that the visual axes are rendered parallel, or within a few degrees of parallelism, by means of accurate surgery. If there is a vertical deviation in addition to the horizontal deviation, this of course must be adequately overcome also.

Much time is often wasted arguing about the precise state of the binocular vision, when it should be appreciated that if a child has once possessed binocular single vision and has developed his binocular reflexes normally, it should be possible to restore these functions if proper conditions are provided.

The more difficult cases are those in which the squint dates from birth or from a very early age, but even in these cases it is often possible to obtain a functional cure by means of accurate surgery carried out within the first two or three years of life.

Yours faithfully,

T. KEITH LYLE.

42, CHARLES STREET, W.1
August 23, 1949.

TRAUMATIC OR "CONCUSSION" CHRONIC GLAUCOMA

To the Editorial Committee of

THE BRITISH JOURNAL OF OPHTHALMOLOGY

DEAR SIRS,—I wish to refer to the carefully-written paper on the above subject by Mr. Arthur d'Ombrain in the August number of this journal (1949, **33**, 495).

The thesis that unilateral chronic glaucoma may be due to previous trauma appears to be founded mainly upon two facts:—

(1) the history of trauma involving an eye, *e.g.*, as a "black-eye," (2) the length of time during which the second eye has remained free from signs or symptoms. I venture to suggest that this is rather a flimsy structure on which to build an edifice so important from the medico-legal aspect. I would draw attention (1) to the fact that the three patients reported were males, and that it must be rather a rare event for a male to pass through life without receiving a "black-eye." (2) The period that elapsed with the patients under observation was 6, 3½ and less than 2 years respectively, since the discovery of chronic glaucoma. It must be the experience of many ophthalmic surgeons to have noted the onset of straight primary glaucoma in the second eye at a much longer interval after its discovery in the first, in patients under regular periodical examination.

Without going through the records of all my private cases of glaucoma, I recall two private patients in this category in whom the interval was 5 years and 10 years, the former male and the latter female. I have little doubt that the former would admit on enquiry to having had a blow on one or both eyes, but I should not on this account classify him as a case of "concussion" glaucoma.

I have no wish to dispute the value of Mr. d'Ombrain's argument. I do, however, feel that the great rarity of the occurrence to which he draws attention should be emphasised, chiefly on account of the medico-legal implications. My view is supported by the "brief and scattered allusions (in the literature) to the possibility of such a lesion" (p. 499).

Yours faithfully,

HUMPHREY NEAME.

149, HARLEY STREET, W.1
August 12, 1949.

NOTES

Prevention of Blindness in Mexico

THE Society for the Prevention of Blindness in Mexico will be celebrating its 4th Biennial Congress from November 6 to November 12, 1949.

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University of Glasgow Department of Ophthalmology Autumn Term, 1949

DURING October a series of meetings will be held in the Department on Wednesdays at 8 p.m. The general arrangements will be similar to those of the series held last year. A discussion will follow the main paper. ***October 5***, Dr. Geo. Leaf—"Biochemical Aspects of Methanol Poisoning"; ***October 12***, Dr. W. O. G. Taylor—"Control of Clotting in Ophthalmology"; ***October 19***, Dr. R. Leishman—"Tobacco Amblyopia"; ***October 26***, Dr. A. Wright Thomson—"Gyrate Atrophy of the Choroid."